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NEUROLOGICAL BULLETIN

CLINICAL STUDIES OF NERVOUS AND MENTAL DISEASES IN THE NEUROLOGICAL DEPARTMENT OF COLUMBIA UNIVERSITY

EDITED BY FREDERICK TILNEY, M.D., PH.D.

VOLUME III
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FOREWORD

THE stress of the past several years has made difficult the continued publication of the Neurological Bulletin along the lines of its original purpose. It has been felt, however, especially by the entire group of its contributors, that the opportunity which this publication offered provided a stimulus in the development of neurology which had a distinctive place in its field. So keenly was the desire recognized by the relatively large group of contributors, that the editors and editorial board felt it incumbent upon them to make every effort toward resuming publication.

The Neurological Bulletin was established under the auspices of the Alumni Association of the College of Physicians and Surgeons in Columbia University; and now again, through the kind offices of Prof. Francis Carter Wood, the Alumni Association has decided to lend its generous financial support and thus assure the resumption of publication.

The policy of the Neurological Bulletin will continue as before, having as its chief aim the detailed presentation of clinical cases, and in all instances following a set and uniform scheme. Articles of general interest will also be included, such, for example, as the leading paper in this first issue of Volume III on "Clinical Endocrinology" by Dr. Walter Timme. It will be the endeavor of the editors to extend their interests into every field which has a direct or collateral bearing upon neurology and psychiatry. Special emphasis will be laid upon the discussions of cases presented at the neurological conferences in Columbia University and the New York Post-Graduate School and Hospital. Oftentimes expressions of personal opinion, especially where divergent interpretations may be put upon a series of clinical observations and facts, form the most stimulating and helpful type of clinical instruction.

The former editor takes great pleasure in announcing that he has associated with himself as editors, Prof. Charles A. Elsberg, to supervise the neuro-surgical work of the Bulletin, Prof. Louis Casamajor, who will direct the neuro-pathological work, and Prof. Thomas W. Salmon, whose interest in mental hygiene and psychiatry will insure the broadest approach to these subjects. The editorial board will comprise Drs. S. Philip Goodhart, Michael Osnato, Charles A. McKendree, Bernard Glueck, Oliver S. Strong, Henry A. Riley, Hubert S. Howe, I. S. Wechsler and George A. Blakeslee.

Frederick Tilney

NEUROLOGICAL BULLETIN

VOLUME III JANUARY, 1921 NUMBER 1

CLINICAL ENDOCRINOLOGY*

By WALTER TIMME, M.D.
Neurological Institute
NEW YORK CITY

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INTRODUCTION

In presenting a moderately comprehensive view of the functions of the endocrine glands, it may be well to divide these organs into groups depending upon their chronological periods of activity. For this purpose, the human life cycle falls naturally into three spans; that from birth to puberty; the next from puberty to the prime; and the last through a gradual deterioration to dissolution. These divisions are not artificial ones, but conform normally to quite marked and certain changes in the internal glandular mechanism. Biologically, there is good reason for such distinction. The first period is that during which there is accretion and growth and development—a gradual process which reaches its acme at a time when the individual has ceased to need the protection of parents and is equipped to begin his independent life. This is ushered in by the full development, physically and functionally, of the organs of reproduction; and concomitantly, by the physical stature and development, alertness and coordination of mind and body which fit the organism to maintain itself against destructive agencies and to enable it to sustain itself in a world of difficulties and obstacles. In short, this second stage of our existence is our raison d'être—it represents the period of advance and aggression and the reproduction of our kind. This accomplished, there is a gradually diminishing necessity for our existence and our final evolutional stage is ushered in.

---

*Lecture to the Fourth Year Students of Medicine at the College of Physicians and Surgeons, New York City

In each of these periods, the goal is different—hence the necessity of a difference in equipment. A lack of equipment in any of the stages, or a gross lack of unity or precision in the arrival of the different factors at any of the critical moments of the struggle, determines the failure of the individual in the competition.

It is our problem to investigate this equipment and see wherein such failure consists; the results of such failure upon the organism, and if possible, its prevention and remedy.

The glandular elements which dominate these stages are the thymus and pineal specifically during the first period, the gonads during the second; with control during both periods exerted in the direction of periodicity, intensity and regularity of the automatic vital functions, by the thyroid, the pituitary and the adrenals. During the final period, there is a gradual diminution in the activity of this latter group up to the cessation of all vegetative functions.

To take these glandular units in order and describe the effect on the body tissues and organs and upon metabolism of their activities, is part of our present aim; but also, in addition, shall we attempt to correlate their activities and to describe their mutually balancing nature and functions so that many complex situations may more readily be interpreted.

THE THYMUS GLAND

EXPERIMENTAL FINDINGS.—The thymus gland has awakened much controversy. From seemingly authentic and well conducted and balanced experimentation, both as to feeding of the gland and as to its extirpation, many investigators have arrived at diametrically opposed views. It had been generally supposed, following the work of Klose and Vogt,[10] that ablation of the thymus gland produced disturbances in the bony structure and growth of the skeleton. Dogs from whom the thymus had been removed showed distinct signs of rachitic skeletal disease. They became bowlegged, had a waddling gait, and could with difficulty maintain themselves erect. The calcium metabolism was likewise deficient and the growth of the long bones was inhibited. In direct opposition to this view, Park and McClure [13] less than a year ago, published the results of their work on thymectomy in dogs, in which, after painstaking care in operative procedure and after-control and operation of their animals, they came to the conclusion that the thymus is absolutely non-essential to life, that its removal has little or no effect on the skeleton or on rachitic disease, although perhaps retardation of growth cannot be entirely denied; that its removal had very little effect on the intelligence of the animal; and perhaps as to its removal-effect upon the other internal secretory glands it could not be absolutely gainsaid that the suprarenals did not become hypertrophied. They denied that thymectomy affected secondarily the thyroid. They declared that the rachitic, bowlegged and waddling dog that was described by Klose and Vogt as a result of thymectomy, was simply in such a condition as a result of the small and low cage in which the dog was confined!

As for feeding experiments, Gudernatsch [4] found that tadpoles were not checked in their growth, but were delayed in their differentiation; while other investigators found that there was delayed closing of the epiphyses of the long bones. Uhlenhuth [21] found that the differentiation was delayed by feeding thymus alone, but this was not the case if other food was given with the thymus. Noel Paton [14] finds that the removal of the thymus and testicles in guinea pigs retards growth enormously, but that removal of the thymus alone does not invariably produce this inhibition. We personally have found, in our feeding experiments upon human beings, that thymus fed to adolescents rarely produces growth unless it is combined with pituitary or thyroid feeding. When so combined, it will almost invariably produce growth in the long bones to a greater degree than when either is fed alone. It seems to us that the discrepancies in the results of extirpation and feeding experiments are not to be credited to the scientific shortcomings of the different investigators, but to the fact that the other endocrine assets of the operated animals vary and cannot be controlled.

From the standpoint of an unbiased observer, it is perhaps fair at this stage to assume that under the influence of the thymus gland—the other glands of internal secretion being normal—the body takes on growth and accretion and that certain inhibitions, chiefly in the direction of the gonads, are at the same time operative. Translated into clinical signs, it means that the marks of sex differentiation are held in abeyance, the secondary sex characteristics being absent.

THYMIC SUBINVOLUTION.—If this interpretation be correct, then an abnormal activity of the thymus gland should be accompanied by certain signs of disturbance in bodily growth and sex differentiation. We see many patients that do show such disturbance and we regard them as "thymic" cases, —with what justice and acuracy will doubtless appear in the near future. These cases may be divided into two groups—one depending for its symptomatology upon a thymus gland which remains active beyond the time when it should normally cease, and the other giving a picture produced by too early a cessation of thymic activity. The first group-picture, termed "thymic subinvolution" has certain characteristics, clinically, which are of importance. These are, as one would suppose, dependent upon a retarded differentiation and upon a continuous skeletal growth. The patients exhibit the following:

1. The skin is of soft texture, smooth and velvety, with a fine covering of lanugo, and a faint or pronounced blush on the cheeks, of a typically "peaches and cream" character.

2. In the male there is no mustache or beard, or if he is over twenty-one, perhaps he must shave occasionally only—once or twice a week.

3. The pubic hair likewise is sparse in both sexes, and in the male it assumes the female characteristics of distribution, namely, a horizontal upper margin.

4. The teeth are bluish white in color and rather lack uniformity in size and development so that a well marked case will present central incisors much larger than usual, with

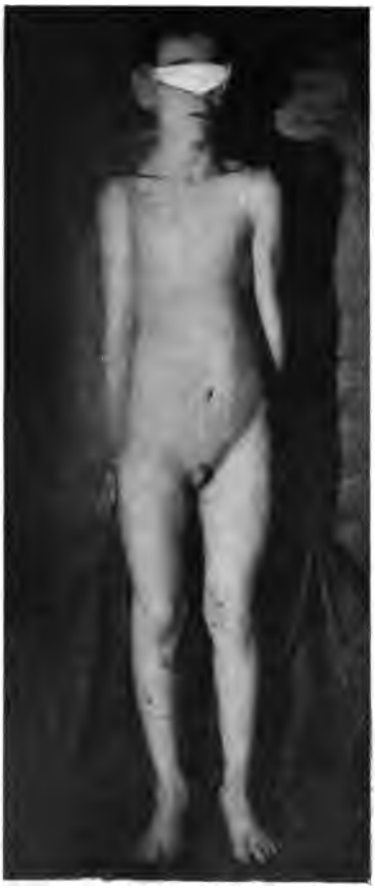

FIG. 1. BOY OF ELEVEN YEARS, SHOWING EXTERNAL EVIDENCES OF THYMIC SUBINVOLUTION. Shows abnormal length of thorax compared with legs. Thighs especially short compared with lower leg. Small genital development with a scrotal fold surrounding base of penis. Arms flail-like. Has large thymus and enclosed sella turcica. Intelligence quotient 0.50. Low carbon dioxide tension of blood. Blood pressure 85:70. White adrenal line. Spasmophilia.

small or undeveloped, or perhaps even absent, lateral incisors.

5. The canines show a lack of normal fang-like character, the edge being cutting like that of the incisors and not pointed.

6. The joint-elements are not firmly bound together so that they are readily hyperextended, even to the point of subluxation in such joints as the mandibular, the shoulder and the metacarpal. The extremities hence give the appearance of flails and the body may frequently be contorted in the most grotesque manner.

7. There is usually present also a marked maxillary torus—the so-called "torus palatinus" at the line of juncture between the superior maxillae in the roof of the mouth.

8. Usually there is delay in the joining of the epiphyses to the shafts of the bones.

Theoretically, the thymus secretes a substance which has vagotonic properties, and practically this is frequently found to be borne out. So that *persistent thymic states are accompanied by low blood pressures, slow pulse, hyperacidity, fatigability, and usually subnormal temperature. The genital apparatus is usually small, but besides this it also shows some lack of differentiation, so that the penis emerges from a scrotal fold which may entirely surround its base.*

Occasionally also, hypospadias is present to a marked degree. In the female, the clitoris is large and projects from the labia more or less prominently. The blood picture frequently shows a moderate relative lymphocytosis, a slight eosinophilia and a low carbon dioxide tension of the blood plasma. The vascular system is somewhat deficient, the arteries being thin and small and the suprarenal glands usually are found small and inefficient. As a result of all these factors, the patients are not sufficiently resistant to diseases and conditions which specifically attack the adrenals, such as diphtheria, scarlatina and influenza. Likewise, as there is a low carbon dioxide tension, the tendency to acidosis is marked. Chloroform anaesthesia, violent shock or emotion, a rapid call upon the adrenalin reserve through sudden excessive activity, frequently bring on collapse and death, without there having been any antecedent cardiac lesion demonstrable. It had been believed that these "thymic" deaths were due to pressure by the enlarged thymus upon the trachea, and although this may often be the case, yet a few autopsies after such deaths have shown hemorrhage into the small adrenal bodies, which accounted for the lethal termination. There is also a lack of tone of the smooth muscle fiber, so that visceroptosis, enuresis and allied conditions are frequent.

COMPENSATORY DISTURBANCES IN THE THYROID, PITUITARY AND ADRENAL GLANDS. —From the foregoing, it might be thought to be a comparatively simple matter to recognize these cases at a glance, but the difficulty is that whenever in the human economy conditions are present that might tend toward the destruction of the individual, compensations occur to offset this tendency. Hence other structures whose secretions are antagonistic to the thymus and its vagotonic effects are presumably called upon to perform such compensation. Such are the adrenals, the pituitary and probably at times the thyroid—all of which have sympathicotonic properties. As a result, and depending upon which of these subsidiary factors is involved to the greatest extent, will we have added to our clinical picture the characteristics of such secondary involvement. This means that signs of dysthryoidism, or dys-

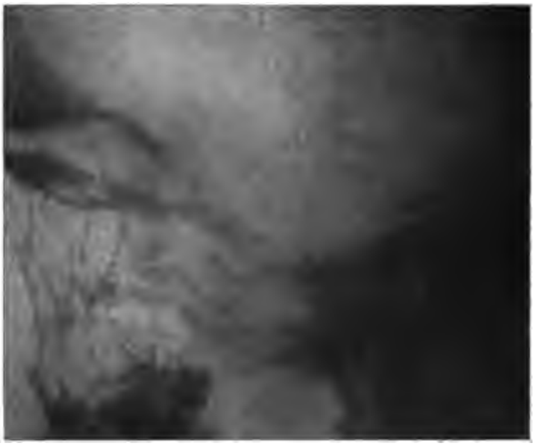

FIG. 2. SELLA TURCICA OF FIG. 1; small and entirely enclosed and "roofed in."

pituitarism or a disturbed adrenal system become engrafted upon the picture. Not to anticipate such characteristic signs, it may nevertheless be wise simply to indicate a few of these superimposed characteristics. The deficient hairy growth may gradually become normal or even increased in amount; the blood pressure become too high and pituitary headaches may supervene. It therefore becomes of the utmost importance, in integrating the symptoms shown by the patient, to determine the chronological order of appearance of the various components of the picture, in order that a proper survey and a comprehensive analysis of the process may be made.

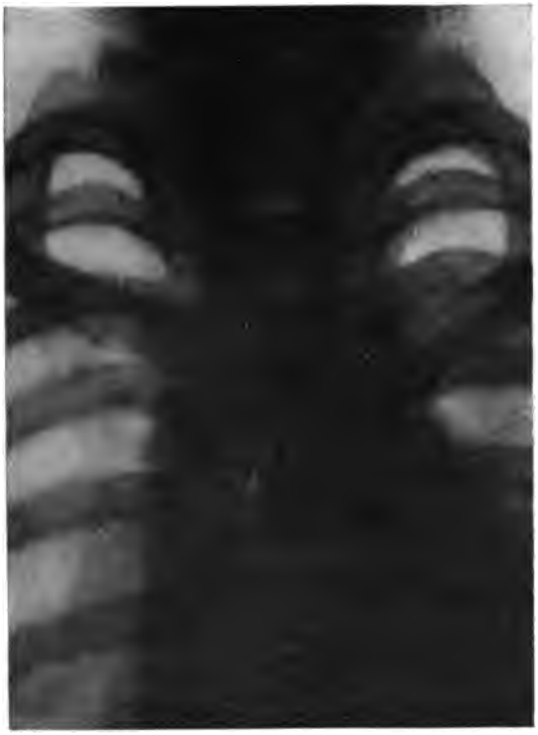

FIG. 3. SHOWS THYMIC ENLARGEMENT OF PATIENT IN FIG. 1.

PSYCHIC MAKE-UP DUE TO SUBINVOLUTED THYMUS.—The mental picture presented by these subinvoluted thymic states is also of great importance, for analagous to their structural lack of differentiation is their psychic make-up. They remain childlike in their character, so that they are self-centered; simple in their mental processes, and imitative; looking for protection and care, and more or less unfitted for the active struggles of life. They are obstinate and negativistic. If, however, an efficient compensation takes place, then, although the mental development may have been delayed, it nevertheless seems finally to reach complete maturity; and these individuals are among the brightest and most intelligent of their community.

PRECOCIOUS INVOLUTION OF THE THYMUS.—In contradistinction to these hyperthymic states, are those produced in individuals by a precocious involution of the thymus. Here we have too rapid a differentiation and as a result the old-young type is produced. The individuals in this group are short in stature, the epiphyses unite early with the shaft of the bone, and with this, the characteristics of an adult are superimposed. Thus, the permanent teeth come in early, though frequently in an anomalous manner. I have seen a lack of one or the other canine, one or both lateral incisors missing, and frequently all the teeth are out of alignment. The secondary hair is early in appearance, pubic hair developing as early as the eighth or ninth year, with development of breasts and a precocious menstruation. One case of this type has come under my observation in which the menses came on in the second year of life—in a mulatto woman of Virginia. These patients are stockily built, with compact frame and frequently vigorous. Their blood pressures are too high, and their blood picture shows an adult polymorphonuclear count even in their childhood days. The genitals are normal in size and character. Of chief interest is their mentality. They are precocious, with much initiative, are easily aroused to anger and are resentful. They have cruel instincts and show little inhibition. Although they seem far advanced for their years while still young, yet they never seem thoroughly to mature and become blocked in early adolescence. They seem to retain their impulsive, unreasoning characteristics, brook no restraint, and remain constantly a prey to their easily aroused anger.

PINEAL GLAND

EXPERIMENTAL EVIDENCE.—Up to very recent times, the question of the glandular character of this epiphyseal appendage was debatable. The older conception of its being the vestigial remains of an amphibian or reptilian parietal eye has practically been decided by the work of Tilney and Warren[19] in the negative, and further than that, the positive evidence of the presence of an epithelial secreting tissue presented by the same authors, gives a basis for the contention that this structure is of the ductless gland group. The experimental evidence produced by McCord[12] and Horrax[6] and to a lesser degree the clinical findings of Timme[20] seem to bear out this supposition. For McCord[12] gives detailed descriptions of the changes that take place in his tadpoles when fed pineal substance, chief among which is the rapid change of their color from opaque olive-green to a transparent, almost colorless vitreous character. Horrax,[6] from his extirpation experiments upon animals, showed that removal of the pineal gland resulted in rapid genital development and growth, and feeding of pineal produced growth and strength of body. Timme[20] finds that early involution of the pineal—as evinced by shadows in the x-ray pictures of the skull—almost invariably accompanies muscular metabolic disturbances, so that fatigability and inability in the muscular system are present. Indeed these disturbances may reach the degree of typical muscular dystrophy, in a number of cases in which early pineal shadows are seen in the roentgenograms. Again here do we find varieties of mental disturbances accompanying the glandular dysfunction.

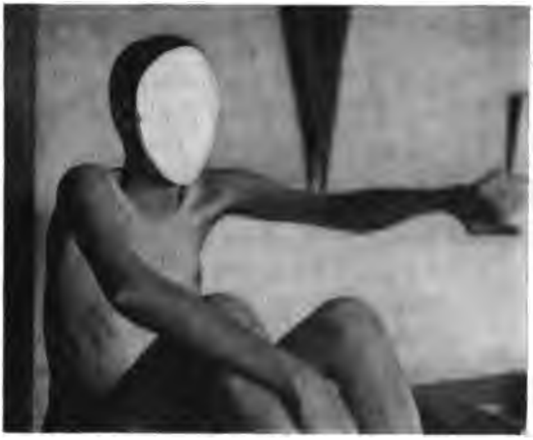

FIG. 4. PROGRESSIVE MUSCULAR DYSTROPHY WITH MARKED PINEAL SHADOW; pineal shadow seen in Fig. 6. Atrophy and dystrophy of muscles are plainly illustrated, especially those at the base of the neck; one bundle of the left pectoralis stands out prominently.

PINEAL SUBINVOLUTIONS.—In cases of subinvolution of the gland, we are confronted with a symptomatology that at the present time cannot be definitely assigned, for the reason that no method has as yet been presented that will give us direct knowledge of subinvolution. When, however, a case presents itself that has the characteristics of a retarded sexual development, both in the genital sphere and in the psychic domain, such retardation being of a direct growth character without the concomitant of reversion or lack of differentiation, pineal subinvolution must be thought of. If, furthermore, the muscular system of such a patient shows undue development and his mental reactions are childish, it is probable that pineal subinvolution is present. Here comes in the theory that pineal activity holds in abeyance pituitary evolution; and hence that the hypopituitarism thus resulting gives rise to the symptoms. Discussion of this situation will take us too far afield and would prove profitless at this juncture. Any change in the chronology of normal glandular function must of necessity cause changes in all internal glands. Even so, the beginnings of such a universal rearrangement presumably have their signs for us to read.

PRECOCIOUS PINEAL INVOLUTION.—The matter lies a little differently with precocious pineal involution, for here we not infrequently have evidence of an early deposit of pineal sand by means of radiography. Normally, such evidence may be gleaned at any time after the thirtieth year. But if pineal shadows are seen in comparatively early life, before the twenties, we have some proof of the early cessation of pineal activity. In these cases we always have a clinical picture differing from the normal. It may be in the genital sphere, the deficient muscular system, or in the mental precocity. Occasionally, though rarely (Fig. 5; dystrophy; large genitals) we see all three fields represented. In this particular photograph, there are well represented the enlarged genitals, and the disturbed muscular system. The muscles have the appearance of those of progressive muscular dystrophy. They also show the contractures seen in this disease as witnessed by the inability to extend the arms completely in all segments. This boy showed a marked pineal shadow. But usually, disturbances in only one field are manifest. There is a blood sugar disturbance—usually one of low content in these precocious pineal cases—analagous to the findings in the myasthenias and in the dystrophies.

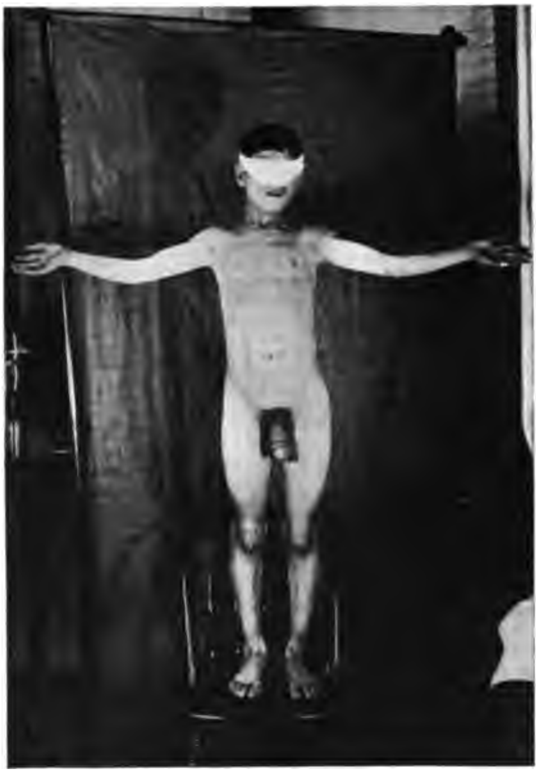

Fig. 5. Boy of Nine Years with Pineal Shadow; showing a combination of disturbances, due probably to pineal involvement, in the muscular and genital spheres. Markedly enlarged genitals; disproportion in thorax and legs; atrophy of the shoulder girdle muscles with contractures in the tendons of the upper extremities preventing full extension of the arms.

If now, we combine the results of either one of the disturbed thymic states with either one of the pineal, we can produce four combinations. It is such a composite picture as any one of these four combinations might show that requires of us interpretation. With each of these four composites we have to add the effect of a similar disturbed function in all the other endocrine organs. The fact needs but to be stated to give some realization of the difficulties, or rather impossibilities, of a correct and demonstrable interpretation of most of the syndromes that greet us.

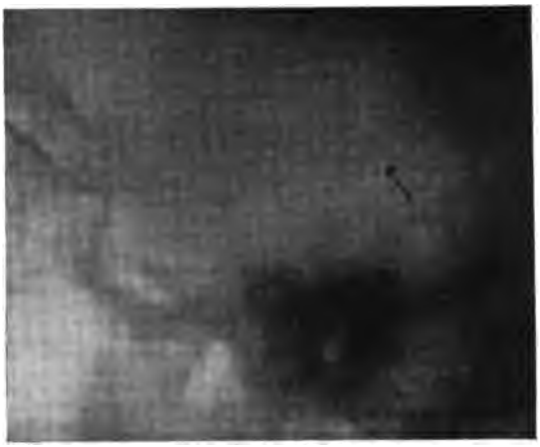

Fig. 6. Skull of Fig. 4, Showing Pineal Shadow.

The Thyroid Adrenal and Pituitary Group.—The next group of endocrine organs that interest us is comprised of the thyroid, the adrenals and the pituitary. It is this group that controls quantitatively and periodically the activity of the other internal secretory organs; and the intensity, regularity and reactive capacity of the functioning of the vegetative nervous system. In short, it controls the automaticity of the individual in his adjustment to environment, in his reaction to fatigue, disease, to anger and other emotions, to competition in the struggle for existence, and prepares the organism physically to meet the shocks attendant upon it.

THE THYROID GLAND

Experimental Evidence.—It is unnecessary here to dilate upon historical allusions in connection with our progressive advance in the knowledge of the activity of the thyroid gland. Suffice it to say that all may be summed up in the work of Kendall[9] of the Mayo Foundation. This investigator has within the past few years succeeded in isolating what seems to be the active principle of the gland in a remarkably high iodine combination. He has designated this compound by the name *thyroxin*. Its experimental administration to animals and men has resulted in the same increased metabolic activity as is produced by feeding whole thyroid gland. And the administration of so small a quantity as 0.001 gram daily has produced profound effects in man; with a rise in the basal metabolism corresponding closely to a proportionate increase of this active principle. The theory of its function in the body is that it acts as a catalyzer—that by its presence the amino-acids which have been produced by partial protein destruction are further reduced by the thyroid principle to carbon dioxide, water and ammonium carbonate; the ammonium carbonate being converted finally into urea by parathyroid action. That is to say, *the thyroid-parathyroid apparatus has for its function the preparation of the results of body oxidation into compounds which can be readily eliminated from the body by the emunctories.* From this we can readily predicate the effect upon the body of disturbed thyroid activity.

Underactivity of the Thyroid.—In underactivity of the thyroid, there are difficulty and delay in the transformation of the amino-acids into lower compounds, and hence the excretory organs cannot properly perform their duty, with the following results:

1. The excretions and secretions generally are diminished. The urine output is lessened, perspiration is diminished, constipation ensues, respiration and the cardiac rate are diminished.

2. The partially decomposed proteins are retained in the tissues, and a diminution of further protein cell destruction follows. As a result, oxidation is lessened, the body temperature is lowered and blood pressure fails.

3. Sluggishness in all domains of activity is the order of the day. The individual suffering from such hypothyroidism reflects all these metabolic changes in his reactions. His body and mind are sluggish, his mental capacity for reception and ratiocination is inadequate; he becomes more or less dull or lethargic and stupid.

4. Tissue change becomes marked. Myxedematous infiltration of the subcutaneous tissue is manifest; skin, nails and hair show the lack of tone and growth in varying degrees depending upon the intensity of the disturbance in the thyroid; and as the lack of metabolic activity is characteristic of all cellular elements in the body, so do we secondarily find sluggishness in the secretory productivity of the other glands of internal secretion.

5. Hypopituitary and hypoadrenal states are engrafted upon an original hypothyroid condition, and a true polyglandular insufficiency is produced.

Individuals of this hypothyroid constitution will vary in their clinical aspects depending upon the chronological advent of their disturbance.

Cretinoid States.—*In early life* such a deficiency produces the *cretinoid states with secondary eunochoid characteristics.* Growth in stature is deficient; hair, nails, skin, teeth show lack of development with but little recuperative and restorative power. Body metabolism is at its lowest point with attendant low temperature, slow pulse, deficient blood pressure. There is a lack of mental development, the victim always appearing of an age below that of puberty, and this characteristic remains if untreated even through several decades. The administration of thyroid substance, however, rapidly ameliorates the condition, although up to certain limits only. The addition of pituitary substance, however, to the therapy, increases its efficiency.

Myxedema.—In later life the disease of the thyroid or general systemic disease during which the thyroid is affected to an underfunctionating degree, or secondary to extensive surgery upon the thyroid, the deficiency produces characteristics in the body which theretofore had been presumably normal. As a result, we get a clinical picture of myxedema rather than one of cretinism, although in certain types of myxedema there is a retrograde process toward a cretinoid state even to the degree of diminution of libido with atrophic changes in the reproductive organs. The mental state also usually suffers degenerative changes. This myxedema however is almost invariably controllable by the administration of thyroid. Autopsy findings have shown that there is an edema of the brain more or less comparable to the myxedematous stage of the subcutaneous tissue.

Intermediate Stages of Thyroid Insufficiency.—Between these cretinoid and myxedematous types, and the normal being, there are numberless stages of thyroid insufficiency. And, differing from these fixed types, the grades of insufficiency do not as a rule remain permanent even without treatment, but are subject to great variations depending upon such factors as menstruation, pregnancy, menopause, seasonal changes, intercurrent diseases and infections. This variability is conducive to much confusion, for if it is not properly understood, an examination of the patient may disclose a seemingly normal thyroid activity due to incidental causes but on a real basis of hypothyroidism. And so arise those clinical pictures in which during structural life, as a result of hypothyroidism, deficiency in hairy growth, poor tooth development, undeveloped sex organs and so on, have been produced, remaining to confuse us at a later day when, although these stigmata of hypothyroidism are present, yet the patient may have become normal as to thyroid efficiency. With this reservation, then, the typical hypothyroid case presents a picture in which all metabolic processes are under par, and this means that every tissue and organ shows the manifestations of this underactivity. The skin is thickened and infiltrated, dry and scaly, with coarse hair which has a tendency to fall out; the teeth are carious, their enamel soft and non-resistant; the finger nails are brittle, grow very slowly and are cracked and seamed. The cardiovascular system shows a bradycardia, a low blood pressure combined with low general body temperature. The basal metabolism is diminished. The secretions are diminished, very little perspiration, a small output of urine and constipation being the rule. The skin reaction is almost nil. Mentally, the patient is dull, sluggish and with little initiative. He

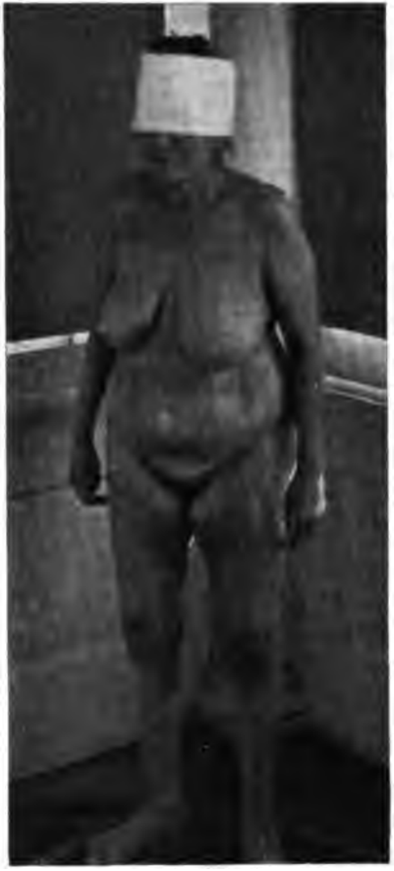

Fig. 7. Case of Scleroderma which has Progressed Half-way Toward Cure on Polyglandular Therapy. Is at present entirely well, with the disappearance of all signs of scleroderma. Illustrates one type of thyroid insufficiency.

moves slowly and thinks slowly, is extremely forgetful, and his lethargy is occasionally disturbed by outbursts of anger due probably to his maladjustment to the more quickly moving world about him. His sleep is profound, and in the morning on awakening much time is required to get into a fit condition for the day's work. While the appetite is usually poor, yet there is distinct gain in body weight, due to the lessened oxidation and output. The external appearance suggests often the picture of cardio-nephritic disease, but the urine shows practically no albumin and an occasional cast. The urea is extremely diminished in quantity. There is a moderate degree of anemia, usually a relative lymphocytosis, the blood sugar is low, the blood sugar curves shows high tolerance and frequently there is a short coagulation time. There are all variants of this picture—some cases of extremely mild type give evidences only in certain domains of their thyroid lack and these are the borderline ones whose economic value can be extremely enhanced by the administration of thyroid gland. It has been frequently shown and confirmed at necropsy that in inefficient thyroid states the pituitary gland partially compensates—the anterior lobe especially being concerned in this process. Hence the reason for the non-comprehensiveness of the hypothyroid picture in many cases. Before leaving the subject of hypothyroidism, however, it is necessary to call attention to the occasional appearance of a goitrous mass even with the hypothyroid state. This is especially so in cretinoid states. These goiters are usually connective tissue masses with some fatty degenerative thyroid tissue and with little or no efficient glandular structure.

In the treatment of this hypothyroid state great care is to be exercised to feed the patients with very low thyroid doses to begin with, and frequently to allow periods of days to intervene between doses. Otherwise the sudden throwing of metabolic end-products into the circulation with overburdening of the emunctories will overwhelm the patient. Indeed, it has been shown by Plummer[15] that the activity of administered thyroid gland is retained for about seven days. Hence the danger of cumulative effect must always be guarded against.

OVERACTIVITY OF THE THYROID.—We are now brought to a consideration of the *overactive thyroid* condition. Again, following out the reasoning based upon the theoretical functions of the thyroid, hyperthyroid states should show the symptoms of an increased metabolism. The partially split up proteins due to cell activity are rapidly transformed by the excessive thyroid secretion and so the path is left clear for more rapid cell change. This produces a rise in temperature, a more rapid pulse, an increase in all the secretions and a loss of body weight. Beyond this, it also causes increased activity in all the other glands of internal secretion so that in truth a polyglandular hyperactivity results. If all these other related structures were synergistic with the thyroid then a condition leading to rapid disorganization of the organism would of necessity result. But certain of these organs are asynergic with the thyroid, notably the gonads and the suprarenal cortex, and therefore a balance is again struck automatically. If these antogonists, however, for some structural or functional reason are inefficient, then the balance is not maintained and the hyperthyroidism is only stayed in its onward course, if at all, by measures designed to limit the cell activity *per se*, or by artificial means of stimulation of the deficient glands or their administration to the patient. A combination of these measures often results in the cessation of the hyperthyroid state. A good example of such hyperthyroidism is that seen in some women after the menopause—the ovarian function having ceased, the thyroid is left without sufficient counterbalance and a hyperthyroid state ensues. This awakens overactivity of the adrenals, with resulting high blood pressure; and the combination produces the flushing, the vasomotor instability, the paresthesiae and so on, of this condition. The administration of ovarian extract or lutein will often allay this hyperthyroidism, cause the cessation of the high blood pressure and the paresthesiae and other aggravating symptoms, and bring about a condition of approximately normal balance. Hyperthyroid states are not of necessity constant. Indeed, their variability frequently is a function of the resultant of the activities projected by the other glands and so a periodicity in hyperthyroidism is usually established. This periodicity is a double one in women—one phase being seasonal and the other menstrual. In spring, hyperthyroidism generally increases. At this point, I should like to state that hyperthyroidism and exophthalmic goiter are not synonymous terms. Hyperthyroidism is a distinct *hyperactivity of the thyroid gland*, and exophthalmic goiter comprises a more or less systemic condition in which the goiter is but a part and represents a *diseased though not necessarily a hyperactive gland*. At the present stage of our knowledge it might even be said that the two states are indicative of different disease processes. As exophthalmic goiter would take us too far afield in this particular discussion, we will confine ourselves strictly to hyperthyroidism as such.

The *causes* of hyperthyroidism are many. They may be classified under the subheads of endogenous and exogenous, although the distinction is at times somewhat fictitious. Endogenous causes would include those that determine endocrine dysfunction, such as status thymico-lymphaticus, under-activity of the pituitary, increased adrenal activity; puberty, or menopause or menstruation; toxins produced in the gastro-intestinal tract, in abscess cavities of the teeth or tonsils and sinuses; too rapid oxidation of the tissues through over-exercise; the ingestion of large amounts of protein containing foods; while among the exogenous factors we find the various infections and contagions, insolations, direct trauma to the gland produced by massage, rough manipulation of the cervical vertebrae, hyperextension of the head in physical exercises, and above all, psychic and emotional traumata such as fright, fear and chagrin. The dysfunctions of the other endocrine units that determine hyperthyroidism are largely produced on an hereditary basis. Hyperthyroid activity has different effects upon the individual, depending upon the age at which the process is inaugurated. If this occurs in the formative period before puberty, lasting structural changes may take place as a result, and even though the hyperthyroid state may later be cured, the structural changes remain. Hence it comes that many patients that show in later years stigmata of hyperthyroidism, are not necessarily hyperthyroid any longer, and medication directed towards such a state is contraindicated. Such signs of an early hyperthyroid state are seen in the rapid early differentiation of the patient; he becomes developed precociously. The gonads reach maturity, the secondary sex characteristics are present, the mental attributes are all acquired early in life. The permanent teeth come in early, attain rather large size and are firm, hard and resistant to infection. There is rapid growth in length of all members, legs, arms and fingers. The bones individually are slender and long. The palate is narrow and high-arched. The thorax is long and narrow and the neck is long. The head is narrow, the eyes usually close together and the cheeks usually rather hollow. The general appearance is one of elongation and attenuation. These are characteristics which remain throughout life and if we are not constantly on guard belie often our conception of the moment.

THE HYPERTHYROID MAKE-UP.—It is not true that "once hyperthyroid, always hyperthyroid." After adolescence is established, puberty has been passed, and the epiphyses are united to the shafts of the bones, the onset of a hyperthyroid state gives a picture which in all its aspects, barring structural differences, is largely the same at all periods of life. Such a hyperthyroid condition may appear in individuals of any and all kinds of body structure—even in those of a hypothyroid make-up. Hence the non-dependence upon body structure in adult life of a diagnosis of a present hyperthyroidism; and hence also, the appearance of hyperthyroidism in a so-called hypothyroid individual. This hyperthyroid picture, so characteristic in all patients, has the following components:

1. A rapid pulse, especially marked and increased after exercise, emotional disturbance, and the ingestion of much protein food.
2. A moderate rise of temperature, the daily variations being from 0.5 degrees to 1.0 degree higher than normal.
3. An increase in the basal metabolism; a progressive loss of weight, with remissions.
4. An inordinate appetite for all kinds of foods, chiefly protein.
5. An increase of all the excretions so that several bowel movements daily are not uncommon.
6. A moderate polyuria is present; perspiration is increased, the body and chiefly the extremities being constantly moist.
7. The finger nails grow rapidly, the beard and mustache likewise. The reaction of the skin to stroking is rapid and productive of a *pink or salmon colored* tache, most marked over the upper chest, but present also even on the arms, abdomen and thighs, which normally do not show such reaction.
8. The *blood pressure* is more or less a function of both thyroid and adrenals and hence will vary in hyperthyroid states depending upon the adrenal reserve. If this is adequate, then the pressure remains fairly high and the vagotonic results of hyperthyroidism do not become manifest. That is, peristalsis is not so rapid, acidity is not increased markedly and fatigue and collapse are not necessary adjuvants as is the case when the adrenals are underfunctionating. The reflexes are exaggerated, spurious ankle clonus frequently being elicitd. There is also a fibrillary tremor of the hands. Muscle spasms are frequent, due probably to a disturbed calcium metabolism.
9. The reaction to the ingestion of glucose is rather prolonged intolerance, shown by the delay in the blood sugar to return to normal. The output of urea is much increased. As a result of the heightened metabolic scale, the individual shows a constant tendency to unrest.
10. Both mind and body are everlastingly busy. And not only with present problems, but anticipatory of to-morrow's as well. The patient knows no rest or relaxation. His mind, filled with echoes of the day's troubles, prevents his falling to sleep until long after he retires, and he is again awake and immediately on the *"qui vive"* as soon as daylight comes.

As a result of the constantly moist skin, the rapid evaporation in draughts of air produces frequent colds and he realizes his weakness in this regard. The quick reaction of the skin is seen in his ready flushing and in the erythematous patches which appear on the neck and upper chest in emotional disturbance. The moist hands gather grime and dust rapidly and hence he washes his hands frequently during the day. This is sometimes one of the first symptoms complained of by the patient or his friends—the constant washing of the hands. In women menstruation is usually irregular in that the periods are too frequent, and the loss of blood is rather profuse with a prolongation to six or eight days of the flowing. Also as a result of the heightened metabolism, unless the emunctories are acting proportionately to the destruction of tissue, there is retention of the oxidized products and cephalagia of a toxic nature results. It must furthermore be remembered that as the increased activity due to hyperthyroidism is present in every cell, the other glands of internal secretion are also involved in this over-activity. It soon becomes evident that the picture is colored with pituitary, adrenal and gonadal touches as a result and at times even a working balance may thus be struck. Thus hyperthyroid women are frequently less so during the week succeeding menstruation, when the corpora lutea secretions are holding sway. The tendency to vagotonia with its attendant low pressure, nausea, hyperacidity and so on is partially counteracted by the increased adrenalin and pituitary output. As before stated, such a hyperthyroidism may be engrafted upon a structure which shows all the signs of an early hypothyroidism or a dyspituitarism or an adrenal dysfunction. If we appreciate this, then the seemingly anomalous combinations become understandable.

SUPRARENAL GLANDS

EXPERIMENTAL AND ANATOMICAL EVIDENCE.—These organs, two in number, are situated deep in the abdominal cavity in a position in which they are both superbly protected and at the same time well placed for rapid efficiency close to the structures they must subserve to maintain life at critical periods. These structures are the large abdominal blood vessels, and the critical periods are those of shock. The suprarenals are composed of two widely different tissues, the cortex and the medulla. The medulla is composed of cells which have an identical origin with the cells of the sympathetic nervous system and are chromaffin in nature. Be it here observed that there are many more collections of chromaffin tissue in the body than are represented in the suprarenal medulla. These are found at the bifurcation of the larger blood vessels—the aorta and carotids for example. Hence experiments to determine the effect of the absence of adrenalin upon the organism that do not exclude these structures from participating in supplying adrenalin, are not to be relied upon without reservations. The suprarenal cortex is embryologically descended from the same tissue as the ovaries and interstitial cells and histologically they are much alike. As a result of their widely different origin, the medulla and cortex of the suprarenals may be looked upon practically as different organs, and indeed, in some of the fishes, the cortex is entirely distinct from the medulla, and forms with its mate of the other side an *interrenal body*. The juxtaposition of cortex and medulla, however, is in all probability purposeful, the secretion of the one possibly balancing that of the other.

Taking up the *adrenal cortex,* our first observation is its relative size compared with the medulla. The ratio in man is about 9:1. In the lower scale of animals, this fraction is much smaller, the cortex being markedly smaller in reference to the medulla. In acerebrate human monsters, the suprarenal cortex has also been found wanting. From these two premises it might be possible to draw a more or less general conclusion that the suprarenal cortex varies directly with the development of the cerebral cortex. Such a conclusion is interesting in its relation to one theory of the function of the suprarenal cortex, namely, that its activity is one of inhibition of the adrenal medulla. We shall see that the secretion of the medulla is of such a nature that it stimulates the sympathetic portion of the vegetative nervous system, that it maintains blood pressure and smooth muscle tone and prepares the individual for the necessary aggressiveness required to survive. But in high development, this animal aggressiveness or combativeness is tempered not only by judgment on the psychic level, but also by a certain inhibition of the vegetative activity occasioned by the adrenal medulla. This inhibitory limitation on the vegetative level is assumed to be applied by the suprarenal cortex. Hence the greater the cortex in relation to the medulla, the higher the organization in the scale of life. The cortex, arising from the same source embryologically as the gonads, would seem to possess properties analogous to them. As a matter of fact, curious changes in the secondary sexual characteristics of the individual are manifested in cortical suprarenal disease. Thus in the female, a disturbed cortex produces a distribution of pubic hair similar to the male distribution, namely, with a triangular upper border, the apex at the umbilicus. And similarly to the testicular and ovarian effect upon blood pressure—namely, a depressive one—so also has the secretion of the cortex a depressive effect upon the medulla with a resultant fall in pressure. Disturbances of the cortex of the suprarenals also result often in abnormal pigment deposits in the skin and mucous surfaces throughout the body, although it is a matter of some doubt as to whether the medulla is not also included in the pathological process.

It has been advanced that the depressor principle in the suprarenal cortex is not an actual secretion but simply cholin, which is a constituent of other tissues in the body likewise, notably the thymus. Addison's disease, which at one time was thought to be due to a suprarenal cortex disturbance, is now generally conceded to involve the medulla as well. Indeed, our knowledge of the suprarenal cortex is very fragmentary, and its real part in the internal economy practically unknown.

The *suprarenal medulla* is on the contrary, the best known of the internal secretory organs, its product, adrenalin, being the first of their active principles to have been isolated. It is now also produced synthetically.

ACTION OF ADRENALIN.—Originally, adrenalin was supposed to be purely a pressor principle. Of late years, due to the researches of Cannon,[1] Hoskins,[6] Hartman [5] and others, it was found to have many effects upon the organism besides that of raising blood pressure. Indeed, even this latter property must be taken within limitations, for in small doses given infrequently, a lowering of the blood pressure has been produced. Furthermore, the change in blood pressure is not universal in all the blood vessels of the body. Upon some systems it is said to have no effect—these are the pulmonary, the cerebrospinal and the coronary arteries. Again the effect is different, and contrary, in the vessels of the skin and of the underlying muscles; contracture in one set accompanying relaxation in the other. This difference in effect, according to Hartman, is dependent upon difference in dosage. *The action of adrenalin is supposed to be exerted on the neuro-muscular synapse, and its action may be enhanced by sensitizing this synapse to adrenalin.* Such is supposed to be the effect of thyroid secretion and cocain. Generally speaking, it may be best to regard the adrenalin as a factor in the maintenance of tone of smooth muscle fiber—not necessarily a contractile spastic activity, but a governing, controlling one, maintaining elastic undulatory conditions. Adrenalin specifically stimulates the sympathetic fibers and ganglia of the vegetative nervous system and hence has a depressive effect upon the autonomic or extended vagus system. The results of this activity are far-reaching. If not balanced, an excessive adrenalin supply will therefore produce the signs of what has been termed *sympathicotonia* including:

1. Delayed peristalsis.
2. Rapidity of pulse.
3. Flushing of the skin.
4. Lessening of the secretions generally.
5. Mobile and frequently large pupils.
6. Deficiency in the gastric juice and in its acidity.
7. Heightened temperature.
8. A heightened blood pressure and a general increase of metabolism.

Such individuals are the active dynamic kind with euphoria, initiative, optimism; easily excited and choleric. One of the specific activities of adrenalin is its effect upon *the mobilization of sugar from the storehouse, the liver.* The researches of Cannon,[1] Langley,[11] Dale [2] and others are in virtual agreement that stimulation of the splanchnics, with normal adrenal glands and liver, produces an increase of sugar in the blood. Stimulation of the splanchnics with the adrenals cut off from their sympathetic influence does not produce this result. It is this added sugar content in the blood which enables the muscles to perform with greater energy than before, and is purposeful in that it fits the individual for the strife or flight which his senses tell him faces him. It is through the intermediary between cerebral cortex and suprarenal glands—namely the sympathetic nervous system—that psyche and soma are correlated in their activities for survival.

ADRENAL INSUFFICIENCY.—In an individual insufficiently equipped with an adrenal apparatus or in whom the relation with the cerebral cortex or other correlated organs is either interfered with or is inadequate, a condition arises which makes such individual unfit to cope with his vital problems to a degree commensurate with such disability. To give concretely the situation determined by this inadequacy is to realize the import of the physiological activity of the adrenals.

There is not a proper maintenance of tone of smooth muscle fiber and hence rapid changes in blood pressure take place with a predominant low systolic reading. Ptoses of the hollow smooth-muscle viscera, with a lack of internal pressure, and loss of rotundity develop.

Hemorrhoidal conditions arise. There is a general loss of capillary tone, so that if the surface of the skin be stroked by the palmar surface of a finger the slight reaction of the vessels immediately concerned serves to distinguish the line thus made from the general surface of the skin and it appears white—the *so-called Sergent line.* An injection of adrenalin just after this stroking will make the line disappear, as will a prior injection prevent its occurrence. This white line is most difficult to obtain in the upper region of the chest immediately in the neighborhood of the thyroid gland, but when it does occur there, it is quite pathognomonic of an inadequate adrenal system, or one that has been exhausted. Such exhaustion may be produced through overactivity beyond the safe limits of the particular individual, through lack of food, exposure to cold, an overbalance of the adrenals by thymus or thyroid or perhaps gonadal activity, and through disease specifically acting upon the adrenals—such as scarlet fever, influenza and a host of other infections. It may be produced by administering too great or too long-continued thyroid medication. Such thyroid medication calls forth the adrenalin through sensitization and the appearance of the white line on the thorax, or a faint pink line surrounded by a border of white is a sign for the discontinuance of such medication. The white line is not necessarily accompanied by low blood pressure—but it always indicates an adrenal inadequacy and the subject is always prone to rapid fatigability and exhaustion. The blood sugar content is diminished and such low readings as 60 milligrams per 100 c.c. of blood are frequently obtained. Many of the myaesthenic conditions show this characteristic, and many of them are produced by the influence of some toxin acting upon the adrenals. The hypoadrenalism may also be a congenital or hereditary state. Status thymico-lymphaticus is usually accompanied by inadequate adrenals. And the external characteristics of the one are similar to those of the other state. The hypoadrenal individual cannot undertake work that requires much energy or requires a sudden demand for exertion or shock. He succumbs to anesthesia, to sudden speeding, to stair-climbing; he is prone to suffer from the various disturbances of a dystonic vascular or smooth muscle system such as spasmodic asthma, urticaria, angioneurotic edema; to vagotonic disturbances such as hyperacidity, gastro-intestinal hypermotility, low carbon dioxide tension of the blood, eosinophilia, intense perspiration, slow pulse, positive oculo-cardiac reflex and the many other symptoms and signs of this syndrome. Many of the so-called "neuro-circulatory asthenia" cases of war-time were of this type. With the lack of the wherewithal to defend or attack, such organisms are the prey of constant fears. Their already deficient adrenals are made still more so by the drain on them produced by their psychoneurotic states of fear. The situation is one of unstable equilibrium and a vicious circle readily establishes itself. Beyond endeavoring to assist the deficient adrenals in emergencies, little can be done for these cases until, through rest and overfeeding, some slight reserve is produced. Then through carefully adjusting exertion to capacity, a slow increase of the limits of muscular activity can be attained, and adjuvants in the nature of pituitary extract can be used as substitutes for the inadequate adrenals. It is inadvisable to feed these patients with adrenal extracts for any great length of time for it seems that such administration retards the activity of their own adrenals. The main dependance is upon alcohol, for this perhaps furnishes a readily oxidizable form of carbohydrate substitute without drawing upon adrenalin, especially if given in small diluted, frequent doses. Coffee, strychnin and other simple stimulants only serve to produce more rapid splanchnic effects upon the already weakened glands and hence can be relied upon only to tide over emergencies. Even here they may be dangerous, if the adrenal exhaustion is complete.

THE HYPERADRENAL MAKE-UP.—In the hyperactive adrenal individuals we see, as before indicated, all the effects of sympathetic overstimulation. This hyperactivity, producing increase in pulse rate, heightened temperature, increase of blood sugar, intense psychomotor activity and a supremely intense individual, is sometimes with extreme difficulty only, differentiated from a hyperthyroid state. In both conditions do we have an increase in basal metabolism, and in both also frequently a tendency to exophthalmos. However, the hyperadrenal state is usually accompanied by heightened blood pressure, whereas the overactive thyroid state is the antithesis in this regard except when accompanied by hyperadrenal conditions. The hyperadrenal condition also produces no such hyperhidrosis as does the hyperthyroid. The red cell count in hyperadrenal states is also usually very high, six or seven million red cells being common. The theory of this polycythemia is that there is greater viscidity of the blood plasma, much of the water being abstracted, and causing thereby a greater concentration of the red cells. The increase is therefore but apparent. It is important to realize the difference in these two conditions, for therapy depends upon such distinction. Other points in the differential bearings are that in the thyroid state, hyperacidity of the gastric juice is present, and there is increased mobility of the intestines with several bowel movements daily. In the adrenal state the opposites are true.

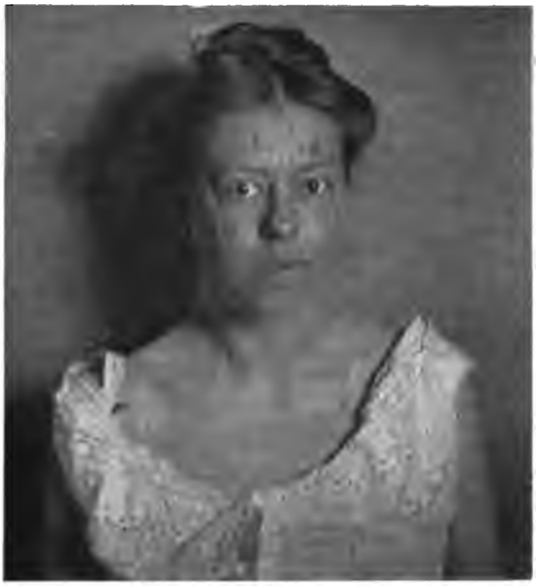

FIG. 8. SEEMINGLY EXOPHTHALMIC GOITER: BUT NO GOITER WAS EVER PRESENT. Blood pressure 180 to 150 systolic; pulse 120; marked exophthalmos; deficient hair on body; outer eyebrow lacking; psoriasis; cessation of menses for one year; no loss of weight; all symptomatic of deficient thyroid activity. When fed on thyroid gland, great rise of blood pressure with rapid exhaustion following. This is a case explained in the text as being due to hyperadrenia and not hyperthyroidism.

THE PITUITARY GLAND

EXPERIMENTAL AND ANATOMICAL EVIDENCE.—The pituitary gland has three distinct parts: (1) The anterior lobe known as the pars anterior or pituitary gland proper, formed by epithelial cells arising originally from the buccal cavity; (2) a posterior lobe, pars nervosa, or the infundibulum, a true glandular formation whose lumen connects with the third ventricle of the brain, and (3) a pars intermedia consisting of an investing epithelium for the infundibulum and divided into two distinct parts—pars tuberalis and pars infundibularis and uniting anteriorly with the pars anterior. The importance of these distinctions is obvious when we know that their functions are different, although as yet only approximately understood, and that the clinical manifestations of their disease are quite different and their therapy correspondingly altered. The anterior lobe in general is believed to control connective tissue growth in the body, especially the skeletal, and to control the activity of the gonads. The posterior is credited with properties stimulating metabolism; maintaining blood pressure not only through the agency of a pressor principle, but also of a depressor one, which has been extracted from the gland with alcohol. It is also said to increase peristalsis and stimulation of the sympathetic system generally, thereby improving nutrition: it increases the production and flow of the mammary glands. It stimulates uterine contractions, increases the urinary excretions, and has lately been credited with increasing the absorption of cerebrospinal fluid. On account of its great sympathetic stimulating properties, it has become the remedy par excellence in surgical shock, being far more efficacious in this regard than adrenalin. The pars intermedia is said to produce a diuretic hormone which is the basis of a diabetes insipidus. The gland as a whole has a direct influence also in genital development, and an inhibitory action upon the activity of the pancreatic juice. The numerous physiological properties of the posterior lobe may be perhaps explained by the result of the investigations of Fühner,[3] who isolated eight different active substances from the posterior lobe. From this short synopsis of the characteristic activities of the hypophysis, in which most observers are agreed, we can readily see to how intense a general disturbance of the bodily functions a disturbed pituitary gland may lead. However, there must be taken into account the fact that the other endocrine glands can partially compensate for deficiency and surplus in pituitary secretion. Thus the thyroid partially compensates for the control and metabolism in connective tissue such as the skin, hair, joints, and so on; while the adrenals partially control through their chromaffin content, the maintenance of blood pressure and tone of smooth muscle fiber; the gonads largely are likewise linked to the pituitary, for they increase and decrease in their activity synchronously with that gland. The pancreas opposes the pituitary, as may be seen by their contrary effect in sugar tolerance.

INTERRELATION OF PITUITARY AND OTHER GLANDS OF INTERNAL SECRETION.—From these considerations, it follows that the cases we see and recognize as pituitary in origin are those in which this compensation by other glands is not entirely sufficient to overcome the original disturbance in the hypophysis—a corollary to which statement is the fact, that, as many, or indeed, all the endocrine glands take part in these various compensatory adjustments, it is extremely difficult and often impossible to determine the original gland at fault; for each gland not only performs the complementary task assigned to it, but produces effects in other directions than just the one that is desired, owing to its changed activity as a whole. The result is then seen in the symptoms produced by this uncompensated excess of the secondarily affected glands which may entirely overshadow the symptoms produced by the originally disturbed gland, and thus lead to a faulty diagnosis. So that, for example, if the adrenals are drawn into compensatory hypoactivity to lessen a blood pressure caused by hyperpituitarism originally, then they will by virtue of this diminished activity also produce pigmentation of the skin. If these two were the only glands disturbed, we should then have a normal blood pressure; but there would be pigmentation as the external evidence of the uncompensated adrenals, and from this pigmentation it would be difficult to diagnose the true condition of a diseased pituitary. Heretofore the cause of such a pigmentation had been assigned solely to adrenal disease, but we can now readily understand how it might be the sole evidence of a pluri-glandular disease originating in the hypophysis. This simple instance will suffice to show what complexity the integration of the clinical manifestations of internal glandular disturbance may assume. Bearing these considerations and strictures in mind, I wish to sketch as succinctly as possible the clinical manifestations evinced in patients with disturbances of the pituitary as the probable prime factor in the pluriglandular syndromes.

ROLE OF THE PITUITARY GLAND IN CERTAIN PLURIGLANDULAR SYNDROMES.—Let us take the extreme cases of disease first as being most easily understood. Here we have, first, the enlargemnt of the entire mass in the sella turcica, due either to tumor or cyst formation with decrease of the pituitary secretions secondary to the pressure exerted upon the hypophysis; and, secondly, a true hyperplasia of the glandular elements with increase of the secretory activity. In either case there appear similar symptoms of direct neighborhood involvement, but with different secondary systemic symptoms, due in the former class of cases to hypopituitary and in the latter to hyperpituitary activity. The direct pressure symptoms give us effects upon the optic chiasm producing bitemporal hemianopsia, (providing that this pressure is exerted anteriorly), that is, blindness upon the temporal side of each visual field, or a primary optic atrophy. The hemianopsia is seen in the early stages for color only and later on for form. If the tumor extends over the edges of the sella, then we get direct pressure symptoms on the sixth cranial nerves, producing paralysis of the external rectus muscles of both eyes, causing internal strabismus; or on the third nerve, causing external strabismus; and still later by backward pressure on the neighboring crura cerebri, the pyramidal tracts are involved, with the resulting disturbances of gait, ankle clonus, and a positive Babinski phenomenon. Of course, such extreme size of the tumor will give intense intracranial pressure with choking of the optic disc, distention of the ventricles, with severe headache accompanied by nausea and vomiting, and, unless relieved by decompressive operations, will result in progressive blindness.

HYPOPITUITARISM.—The systemic symptoms due to the deficient secretion of the hypophysis under such pressure conditions—that is, those due to *hypopituitarism,* are:

1. Increase of body weight.
2. Increased tolerance for sugar.
3. A lowered body temperature.
4. A sluggish mentality with slowness of movement, and with diminution of sexual activity.

If the condition is present in adolescence, and the growth of the tumor rapid, we get infantilism including:

1. An undeveloped sexual growth.
2. Lack of hair, or a tendency toward the other sex in its distribution, and adiposis.
3. In boys, developed mammary glands with general feminine contour.

HYPERPITUITARISM.—Should the patient be seen in the very early stages of the new growth when the tumor is slow growing, at a time when as yet only its irritative effects upon the pituitary gland are exerted, and no actual destruction of the gland has taken place, the syndrome is one of *hyperpituitarism;* and is in this early stage therefore practically the equivalent of a hyperplasia of the gland. Should this hyperplasia arise in early life, before the epiphyses have become ossified and united, then its effect is seen:

1. In the rapid growth of the long skeletal bones, leading to *giantism.*
2. In increased sexual tendencies and development, and precocious puberty with an increased growth of hair.
3. In low sugar tolerance with increased urinary output.

Should the irritating medium arise later in life after the ossification of the epiphyses, then the gigantism does not appear, but we have an abnormal growth effect produced upon the bones of hands and feet instead with resulting *acromegaly.* With the acromegaly is seen the typical skull—a protruding forehead, prognathous lower jaws, wide molars, kyphotic spine; in short, the picture of the deformed court jester—the skull *en polichinelle.* Such people frequently have a highly developed and susceptible autonomic system and are witty and keen. Short of these extremes, we have an infinite number of grades of disturbed pituitary gland with endless combinations and alternations of hypofunction and hyperfunction. These lower grades, as was suggested earlier in this paper, are the important ones to recognize, for only in the prestructural change conditions can we hope to accomplish results with means short of surgical interference.

THE PRE-ADOLESCENT PERIOD.—Let us take up the pre-adolescent period of life. We see a child that presents one or more of the following characteristics; small stature, adiposities, ununited epiphyses, with small sexual organs, perhaps reversive in type, or with cryptorchidism. The skeletal muscles are weak, the teeth malformed, crowded, and irregularly placed, there is a prognathous upper maxillary with a generally deficient bony structure, perhaps even to the extent of acromikria. There may be a weakness of the bladder walls leading to enuresis, with a high sugar tolerance and an abnormal desire for sweets. The boy's temperatment is so different from his fellows' that he can with difficulty get along with them in play; he cries easily and is apt to be cowardly. There may be added a dryness of the skin, subnormal temperature, slow pulse, and low blood pressure. Occasionally there are epileptoid attacks. In girls after puberty, there is apt to be amenorrhea. Any of these symptoms may point to pituitary deficiency, but if several are found combined in one individual, then the presumption is strong that such a deficiency exists. In the other type of case, hyperpituitary in character, the child is a large, spare, and bony framed individual, eyes wide apart, broad face and large feet and hands, a square protruding chin, with early profuse growth of axillary and pubic hair and perhaps even a general hypertrichosis, with rather thick skin and large sexual organs. There is precocious puberty. The teeth are broad, large and unspaced, or normal in size, though spaced, and there may be polyuria with occasional traces of sugar in the urine. The temperament of such a child, unless the process has gone too far, is one of aggressiveness, of pugnacious tendency. Later, should the hyperfunction continue, his mind becomes sluggish, he suffers with headaches, and the more marked structural changes take place.

DYSPITUITARISM.—It might be well to add here that the more advanced symptoms are possibly due to the change from early hyperfunction to the later succeeding hypofunction—a frequent sequence. One must always remember that both processes may at different times affect the same individual. Whether intercurrent disease fans the flame which was dying out, or blows and falls cause an irritation of the gland which had become dormant, or yet that the other endocrine glands influence the pituitary, suffice it to say that there are individuals in whom we see mixtures of these clinical manifestations pointing to alternating hyperpituitary and hypopituitary disturbances called *dyspituitarism.*

In this connection we may mention the series of cases with epileptic episodes. It has been stated by Horsley[7] that undue excitability of the cerebral cortex supervenes after posterior lobe excision or in posterior lobe deficiency, which might account for the seizures. My experience has been that these cases are usually dyspituitary in nature, i.e., they give many evidences of a hyposecretion of the gland—dry skin, amenorrhoea, adiposity, malformed teeth, narrowed eye distances—high sugar tolerance, low temperature and pulse rate, combined with the hypersecretory conditions—much hair, high blood pressure, excitability and with excessive sexual irritability. In these epileptic attacks the patients frequently have so-called uncinate fits, due probably to the neighborhood symptoms of an enlarged pituitary influencing the uncinate gyrus of the temporosphenoidal lobe. These patients frequently have dreamy states associated with gustatory and olfactory impressions without true convulsions; and their epileptic aura is usually one of taste or smell disturbance. In dyspituitarism also, we may find a generalized status thymico-lymphaticus with excessive adenoid formation, frequent nose bleeds and intermittent mucous discharges into the pharynx. Now, clinically, we find all conditions and combinations of the foregoing signs and symptoms, and added to them, I have also frequently noticed the abnormal adaptation of the patient to his condition and environment, and in his emotional bearing and control. Usually patients with the clearly defined hyperactive pituitaries show in their early and transitional stages, precocious psychic development. They are self-contained and calculating, exhibiting self restraint in an unusual degree; usually, by quiet poise indicating great latent power, they control the situation in which they find themselves with playmates older than themselves. The hypopituitaric, on the other hand, is mentally and bodily sluggish, dull and apathetic, and backward at school; and he loses control at the slightest difficulty, and is dismayed at the problems he faces. At times he is the victim of compulsion neuroses—pathological liar; kleptomaniac; unmoral.

The importance is in the early recognition of pituitary disturbance. Does the patient that you believe simply to be under par or overstrung, complain of headaches, does he readily get drowsy, has he subnormal temperature, does his nose bleed readily, has he deficient perspiration even on the hottest day, are his eyes too close or too far apart to be normal, is his hairy growth peculiar in any particular, are his teeth abnormal in character or spacing? These, and similar queries indicated herein, that heretofore received but scant attention from us on account of their seeming irrelevance to the condition complained of, are the vanes pointing in the direction whence comes the storm. If you wait until his visual fields are destroyed, or his choked discs cry for relief, or his acromegaly has become a real deformity; then you have lost valuable time in his treatment; your patient is beyond help. The early recognition of pituitary disturbances is equivadent to a reclassification of many disease entities heretofore described as functional.

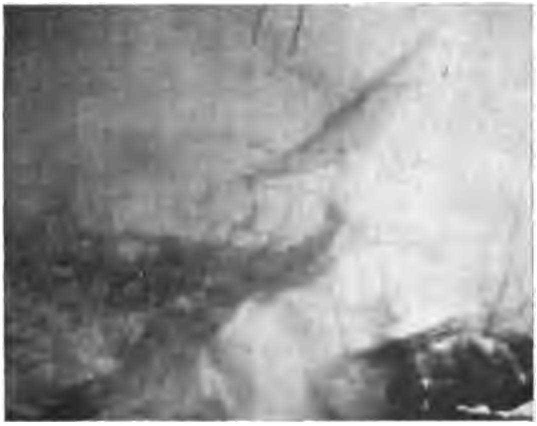

Fig. 9. Showing Sella Turcica with Marked Posterior Erosion, causing free communication between the sphenoid sinus and the pituitary fossa; the clinoids largely closed in by the superior portion of the cavity. The patient had intense headaches for ten years; intratemporal in character; improving automatically as erosion proceeds but more rapidly on pituitary feeding.

Radiographic Types of Sella Turcica.—A few words regarding the radiographs of the skull and the conclusions that may be drawn from the sella turcica outlines. So much has been uttered as to the interpretations that may be made of various so-called deformed sellae turcicae that a few photographs of extreme abnormality are included in this paper. As a general rule, these sella pictures fall into five groups:

1. Generally contracted, with clinoid processes that close in the cavity to some degree, but with no erosions.

2. The same type as 1, but with evidences of erosions in the clinoids or the dorsum or the floor of the cavity.

3. Normal size of sella without erosion.

4. Normal size of sella with evidence of marked erosion.

5. Abnormally large sella with or without erosions.

Types 1 and 2 are pathological and almost invariably produce clinical symptoms—type 1, of simple hypopituitarism in one of its several forms; type 2, of hypopituitarism with pressure symptoms involving the chiasm, or the oculomotor nerves or the tracts in the crura cerebri, with headaches

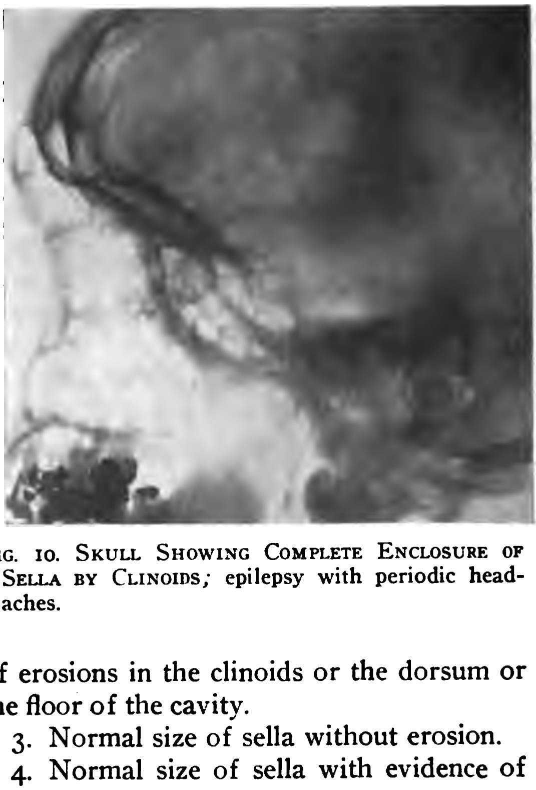

Fig. 10. Skull Showing Complete Enclosure of Sella by Clinoids; epilepsy with periodic headaches.

and perhaps epileptiform seizures. Type 3 admits of no conclusion regarding pituitary disease although pituitary disease cannot be excluded. This type includes by far the largest number of cases. Types 4 and 5 are both pathological and include cases of hyperplasia and tumor or cyst with all the attendant symptoms of general intracranial pressure together with neighborhood pressure symptoms and metabolic disturbances.

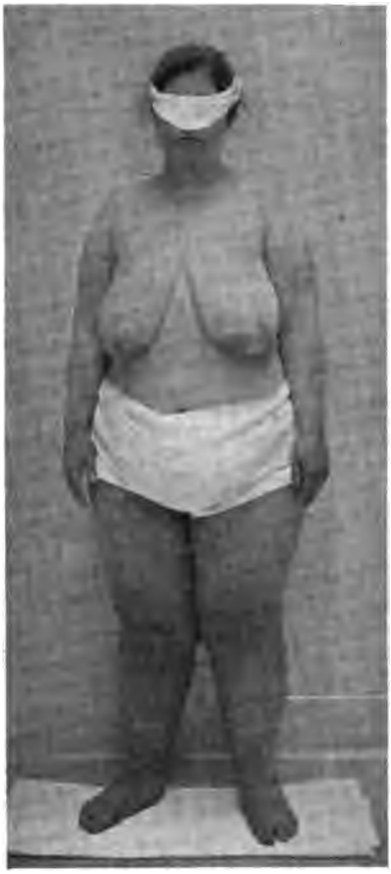

FIG. 11. PITUITARY OBESITY. Girl of nineteen; very small hands, small mouth; adiposity confined to lower segments largely; fed on thyroid injudiciously and collapse ensued. Improved very much on pituitary feeding. Has small enclosed sella turcica.

DIRECT PRESSURE EFFECTS OF SELLAR MASS.—A few words of warning also regarding the regularity of certain symptoms referable to direct pressure of a sellar mass upon surrounding structures, may not be amiss.

In the first place, unless the tumor mass presses directly forwards, ***no hemianopsia will occur.*** Hence the possibility of an hypophyseal tumor without hemianopsia.

If the mass presses posteriorly, then it is possible to have a symptomatology dependent upon the amount of pressure or irritation exerted upon either or both crura cerebri; that is to say, either or both pyramidal tracts may be affected, as well as either or both sensory tracts and the consequent sequelae produced—positive Babinski, exaggerated reflexes with clonus and sensory changes—all

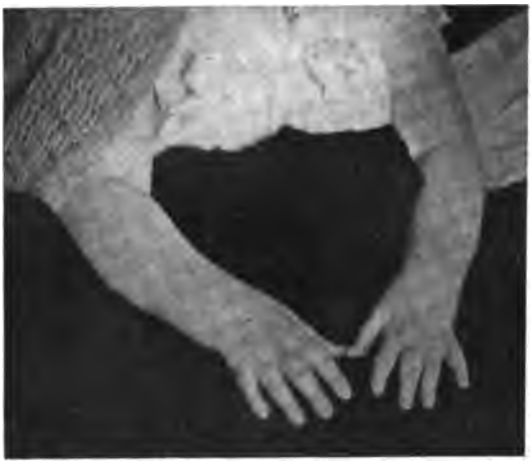

FIG. 12. AKROMIKRIA; Chubby, child-like hands in adult woman weighing 200 pounds, due to hypopituitarism; with very small enclosed sella.

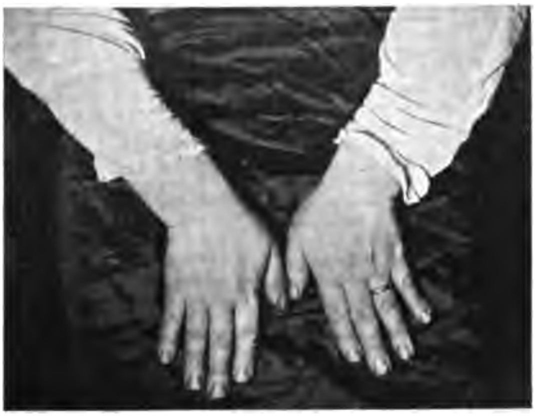

FIG. 13. ACROMEGALIC HANDS; in adult woman, age fifty-three years, whose acromegaly terminated at thirty-fifth year spontaneously; sella very large and eroded.

these without either hemianopsia or oculo-motor palsies.

With a lateral enlargement of the mass, oculo-motor palsies and disturbances of circulation in the eyeball due to pressure upon the cavernous sinus may occur. All of these encroachments of the sellar mass produce headache of the intratemporal type—now known as *pituitary headache*.

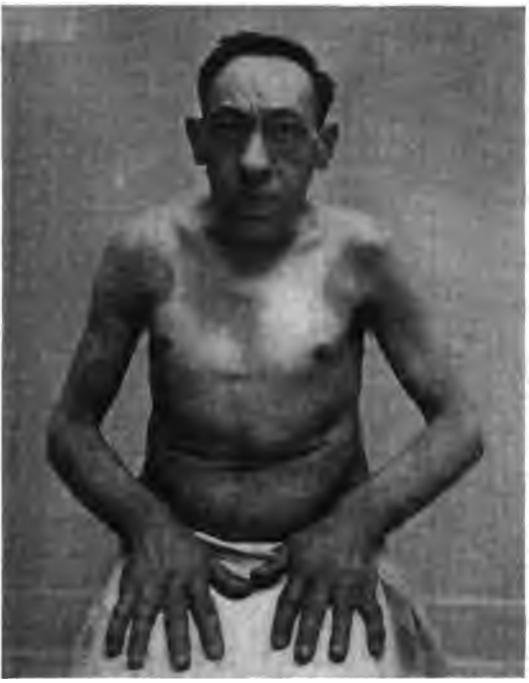

FIG. 14. FAR ADVANCED ACROMEGALIC; pigment patches on shoulders and arms antedated the acromegaly. They followed upon an obscure abdominal affection, probably tuberculous; and are an expression of suprarenal insufficiency. The attempt by the pituitary gland to compensate for the deficient suprarenals possibly accounted for the acromegalic symptoms. Sella was markedly enlarged.

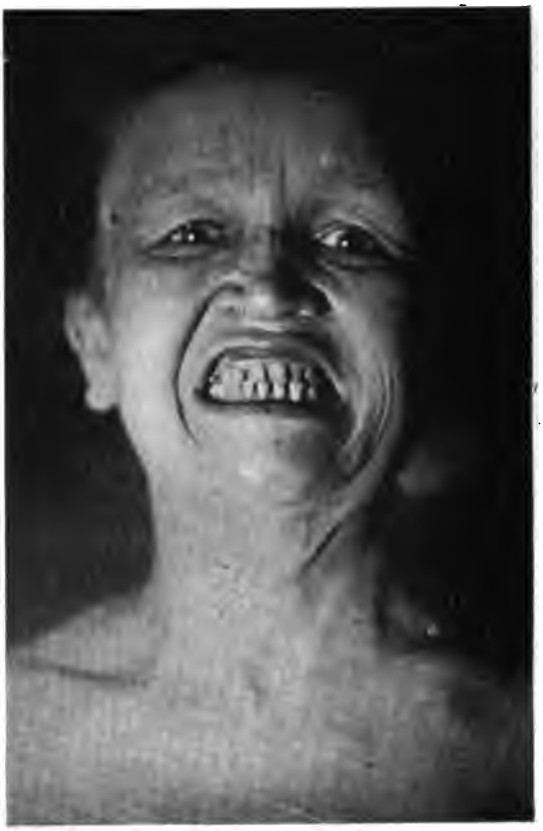

FIG. 15. PAGET'S DISEASE. Confirmed by *x*-ray of bones. Patient shows many endocrine anomalies; almost no eyebrows, and no hair on body. Teeth spaced and stratified; left clavicle shows bony accretions; skull shows very small and enclosed pituitary fossa and practically no frontal sinus; condition much relieved by feeding pituitary and thyroid.

ACTION OF PITUITRIN.—The active principles of both anterior and posterior lobes of the pituitary are now said to have been isolated and at least one of them, that derived from the posterior lobe, known commercially as pituitrin, is of great potency. Its use, especially hypodermatically, has a specificity of action upon the contractile power of smooth muscle fiber so that a heightened blood vessel tone is produced, and thereby an increased blood pressure; uterine tone is similarly increased, so that in cases of hemorrhage through laxity of its walls, in the third stage of labor, in menorrhagia and metrorrhagia, its use is followed by prompt cessation of the flow. I have even seen its careful use in atonic uteri that habitually abort, followed by a cessation of this habit. The atonic bladder wall occasioning nocturnal enuresis, is often much benefited and the annoying symptom alleviated or cured by its use. In shock of all kinds, surgical, anaphylactic and emotional, pituitrin is a valuable measure. In late years, the annoying polyuria of diabetes insipidus has been controlled by hypodermatic injections of pituitrin. In severe cases, pituitrin may be given, diluted with normal saline solution, intravenously. The severe headaches of acromegalics and the bitemporal hemianopsia of pituitary hyperplasia occasionally succumb to its use. It is, however, to be strictly observed, that long continued use of this remedy produces intense asthenia and also that on account of its effect in increasing peripheral resistance, it should not be used in cardiac disturbances with uncertain compensation. The active principle of the anterior lobe is said to have been isolated by Brailsford Robertson[16] and named by him, *tethelin.* He states that it produces all the effects—especially of growth—inherent in the anterior lobe of the pituitary. Sufficient data have as yet hardly accumulated to substantiate this claim.

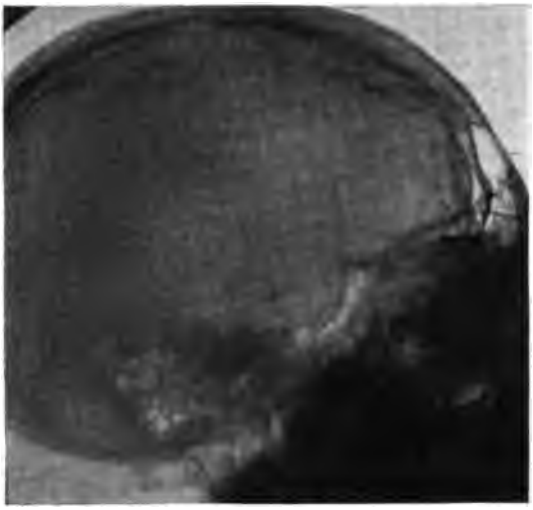

FIG. 16. SKULL SHOWING EXTREMELY LARGE SELLA IN AN ACROMEGALIC, WITH THINNING OF THE WALLS AND EROSION OF THE CLINOIDS. Bitemporal hemianopsia present. Intense headaches intratemporal in situation. Partially relieved by intravenous injections of pituitrin.

THE GONADS

EXPERIMENTAL EVIDENCE.—It remains for us to outline some of the salient features of the *Gonads* and their relation to others of the internal glands. Experiments carried out in this field have been numerous and the results intensely interesting. As outlined earlier in this paper, disturbances and operative experimentation on the thymus and pineal glands have had effects upon gonadal development. Overactive and subinvoluted thymic glands produce in the gonads lack of differentiation so that the male organs approximate the female in that the penis emerges from beneath a scrotal fold which in a measure represents the labia majora. In the female, the clitoris is large and protrudes from the angle of the labia. Pineal early involution brings in its wake early gonadal development with secondary sex characteristics, while delayed involution produces a lack of sex gland development, although providing the thymus is normal, this lack of development is simply

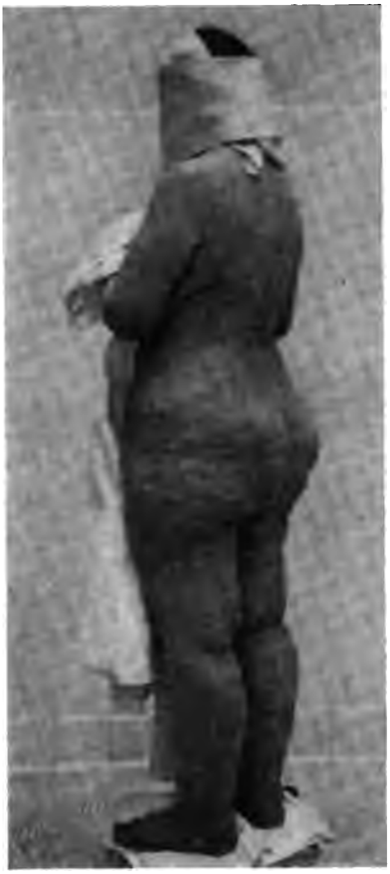

FIG. 17. ADIPOSIS DOLOROSA (DERCUM'S DISEASE). Showing modulated character of adiposities, many of which were extremely painful; largely confined to the lower segments. Followed upon birth of child weighing almost 20 pounds (sic.). The sella turcica was small and enclosed and pituitary feeding ameliorated the condition.

in size and not in character as is the case with a thymic subinvolution. In adolescent and adult human beings with normal gonads, disease of these organs or accidents and injuries often produce a change in the secondary sex characteristics as well, so that we find a change in the voice, disturbances in secondary hairy growth, a change in the contour of the body and frequently also a definite change in the mental characteristics leading to a species of acquired infantilism. The experiments of Steinach,[18] of transplantation of male interstitial tissue into ovariectomized females (guinea pigs) with the attendant subsequent changes of the entire animal into one with male characteristics of growth and hairy distribution, with accentuation of the clitoris; and the similar changes due to transplantation of the ovarian structures into castrated males are striking. Still more suggestive are the experimental researches of Knud Sand,[17] who after castrating both male and female rats and guinea pigs transplanted both ovaries and interstitial male tissue into one animal. This animal took on the characteristics of a true hermaphroditism, with mammary secretion and spermatogenesis. In a litter of males, it comported itself as a female and with fe-

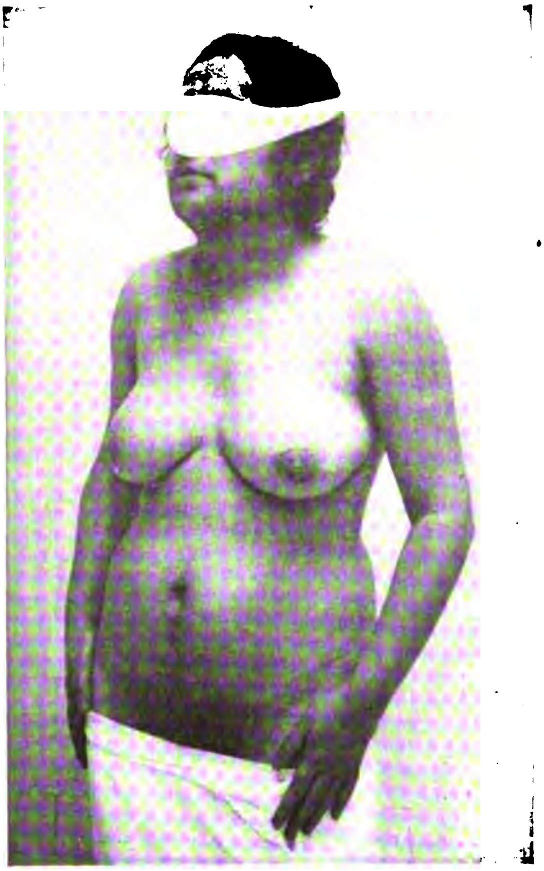

FIG. 18. MASCULINE HAIR DISTRIBUTION IN FEMALE. Mustache, beard, masculine pubic hair; psychoneurotic; menses established at nineteenth year and ceased at thirtieth with intervals of six months between periods. Maladjusted, depressed, shut-in type. Thirty-five years of age.

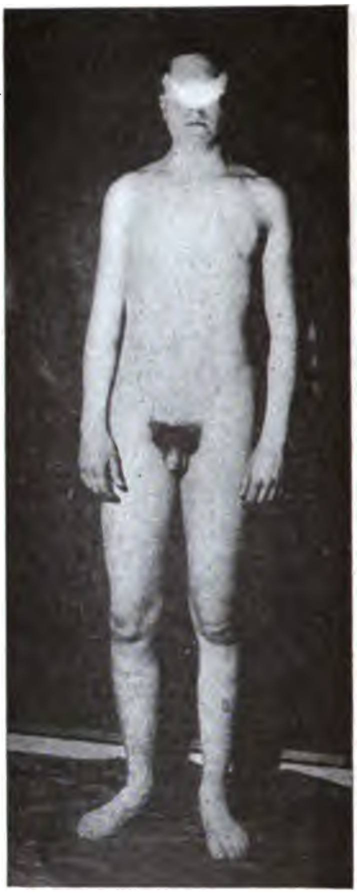

FIG. 19. FEMININE HAIR DISTRIBUTION IN MALE. Never shaved; no hair on body except pubic region; great fatigability; feminine attributes, although he looks powerful and pugnacious and is over 6 feet high. Is occupied as typewriter and stenographer. Psychoneurotic; maladjusted. Low blood pressure, low blood sugar, exhaustibility; also presents a subinvoluted thymus gland and a small but eroded sella turcica.

males as a male. He finally transplanted ovarian tissue directly into the scrotum of the male and really produced thereby a bisexual organ with both corpora lutea and spermatogenetic formation.

RELATION OF GONADS TO OTHER GLANDS. —The relationship between the gonads and the other organs is not fixed. During the period following menstruation when the corpora lutea are formed, there is usually a diminished thyroid activity. As this period of luteal activity begins to wane, and the next menstrual period approaches, the thyroid, presumably less opposed, becomes overactive and the nervousness, excitability, and general mental uneasiness produced by thyroid activity become manifest. This again succumbs to the fresh luteal secretion. However, when, through nonactivity of the ovary—at the menopause for instance—this is not the case, then the persistent thyroid, uncompensated by luteal secretion, produces many of the symptoms of this state. The paresthesiae, sleeplessness, nervousness, irritability, cardiac arrhythmia, vasomotor disturbances and profuse sweating of this condition are well known: and are in all probability due largely to thyroidal disturbance. The increased blood pressure seen frequently at this time is due to the sensitization of the neuro-muscular synapse to adrenalin by the thyroid activity.

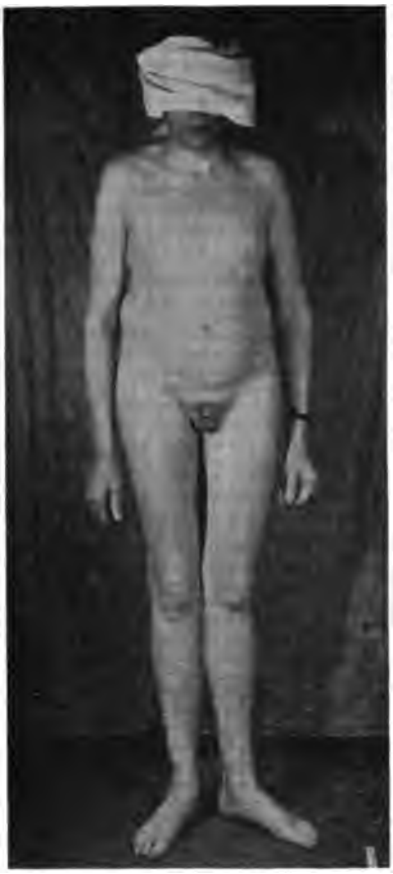

FIG. 20. INFANTILISM. Age forty-one. Voice high-pitched and soft; testicles undescended; moderate mammary gland development. No hair on body except a few sparse hairs on pubes; never shaved; abdomen of feminine type; was 21st child out of 22 children. Skull shows small sella, moderately eroded; thymus gland shadow present in x-ray plate. Is extremely competent in business. Has some phobiae relating to early senility.

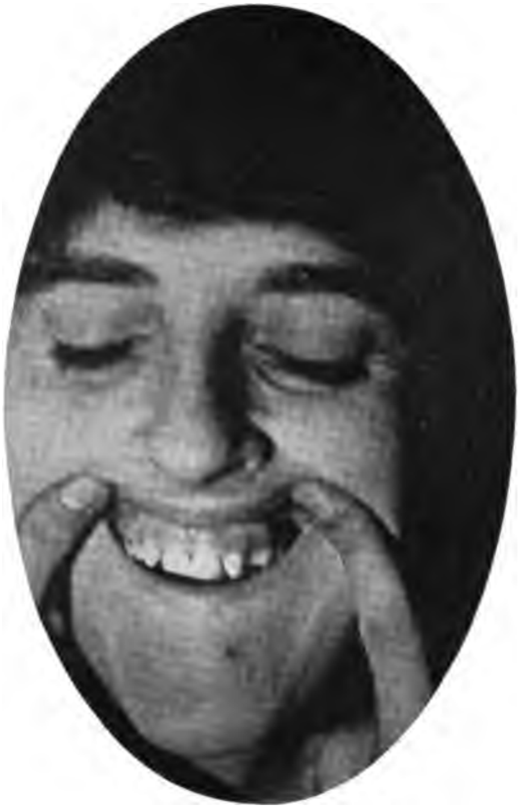

FIG. 21. SHOWING ABSENCE OF LATERAL INCISORS OF THE UPPER JAW OF A YOUNG WOMAN OF TWENTY-FIVE. She has never menstruated; x-ray examination shows no hidden incisors and gynecological examination shows rudimentary uterus and an absence of ovaries.

If this adrenalin content is deficient, then we get intense weakness, asthenia and collapse. The administration of lutein at this time offsets this thyroid activity and often brilliant results follow in the alleviation of the very annoying conditions produced by it. The relation of the thyroid to the gonads is also strikingly shown in the increased rapidity of sexual development under its influence. In amenorrhea, or in delayed and scanty menstruation, thyroid administration is effective. In the delayed differentiation due to thymic subinvolution, with the testicles still in the abdominal cavity, thyroid administration is a potent agent in producing their emergence through the canal into the scrotum. The relation of the pituitary to the gonads needs hardly to be stated. It is universally known that if the gonads be ablated in early life, great stature results, due to a secondary activity in the pituitary gland. Thus the eunuchs. It is also believed that the periodic headaches that accompany menstruation, are due to temporary pituitary hyperplasia. The anterior lobe of the pituitary is believed to be the controlling factor in the gonadal balance. It is a fact in my experience that radiographs of the sella turcica in which the anterior clinoids dip down into the cavity of the sella, contracting it especially in its anterior compartment, are accompanied by malformed or abnormally developed sexual organs and curiously enough by disturbances in the psychosexual sphere. Ablation of the testicles or of the ovaries produces in individuals that have in combination with a hypersexual make-up, qualities of the unmoral imbecile, and that are hence dangerous to the community, a more or less complete docility and they may be thus partially regenerated to become moderately useful economic factors.

Investigators of a decade or so ago advanced the theory that as a result of early sex maturity, the epiphyses became joined to the shafts of the bones earlier than normal and hence growth in stature ceased; conversely, that delayed gonadal development caused a delay in this ossification, with attendant growth in stature. This is true in the author's experience in the majority of cases, but the exceptions are so numerous and so prominent that the theory as a universality must fall. The probable reason underlying the situation is that the growth factors represented by the thymus and pituitary glands already mentioned are properly or improperly balanced and thus produce variations in the growth of the long bones. But both of these factors also determine to some extent gonadal development though in an antagonistic way, whereas in growth of stature their activity is synergistic. Thus various combinations might readily arise in which both gonadal development and stature growth were variously and at times diversely limited in the same individual. Similar failure of so-called universal laws controlling endocrine manifestations rests upon like reasoning.

In closing this lecture, in which of course, only some of the salient points of the vast subject are touched upon, it may not be amiss to outline the dynamic nature and the continuity of the processes in the internal glandular system, which enable the individual to emerge from his inherited difficulties to become a normal economic factor. To do this concretely, let us follow the development of the child that in its early years is the subject of that disturbed glandular situation, "status thymico-lymphaticus." If the disturbance is critical, early death ensues as a result of too sudden and great demand upon its energies—in running, during anesthesia, in emotional stress. But, if spared from these eventualities, one sees a gradual change take place in its constitutional reactions depending upon various internal glandular compensations. It will be remembered that status thymico-lymphaticus is accompanied by low blood pressures, intense fatigability, low blood sugar content, low carbon dioxide coefficient, a low polymorphonuclear count and in general a deficient chromaffin system and an underacting pituitary gland. If there were a possibility of the pituitary becoming overactive, it might compensate for some of the suprarenal shortcomings, such as extreme fatigability, low blood pressure, low blood sugar. This can be accomplished in some cases without any apparent manifestations of the disturbance excepting that of increased growth. But more frequently the pituitary in these status cases is confined within very narrow limits by a small and closed-in sella turcica. In order that enlargement and hyperactivity may occur on the part of the pituitary, this narrow enclosure must give way. As a matter of fact, erosion of the clinoid processes does occur, together with excavation of the cavity in all directions, notably posteriorly and in the floor, even extending into the sphenoid sinus. In this process of enlargement of the sella turcica, however, due to the mechanics of the situation, many symptoms arise. Pressure of the hypophysis against the sella walls produces the intense intratemporal headaches, lasting over a number of years, migrainous in character, of which so many adolescents

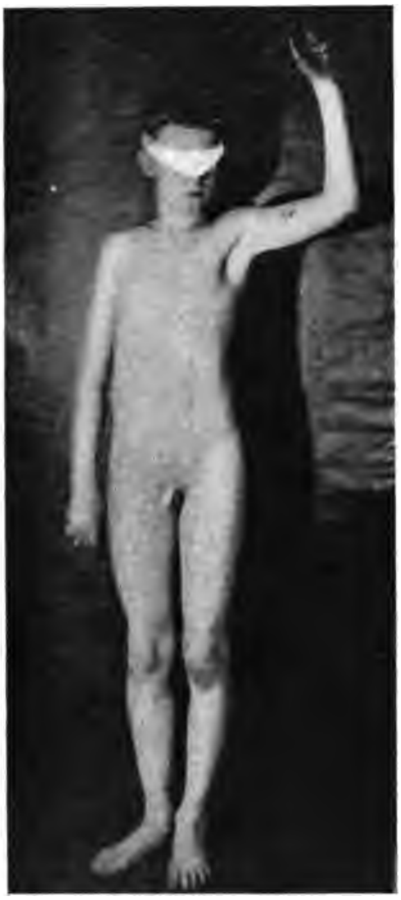

Fig. 22. Case of Status Thymico-lymphaticus Emerging from this Condition as Described in Text. This case has a very small sella, has an undeveloped genital system with scrotal implantation about the base of the penis; has no hair on body; bruises easily as seen from slight pressure of thumb on raised arm a few minutes before picture was taken; has spasms of muscles and enuresis; x-ray shows thymic shadow; low blood sugar, low carbon dioxide tension. Patient is a pathological liar. Age thirteen years.

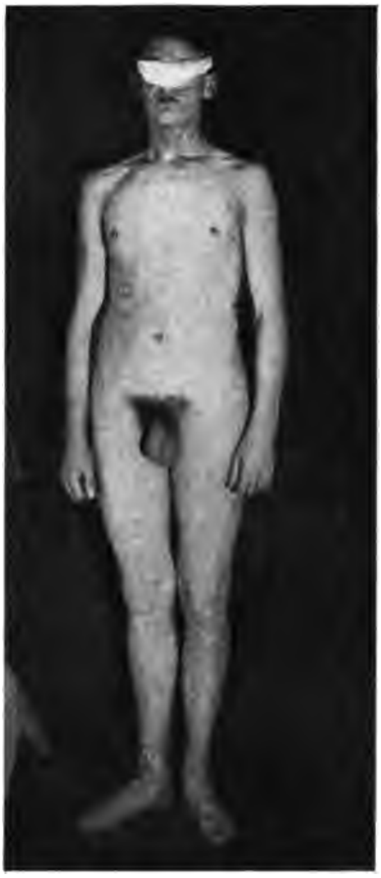

Fig. 23. Later Development of a Case of Status Thymico-lymphaticus. Still shows thymic shadow; sella turcica much eroded; feminine pubic hair; although he looks strong and well-developed yet he has intense asthenia, low blood pressure, low blood sugar and low carbon dioxide tension. Patient over 6 feet; never shaved; twenty years of age; x-ray shows thymic shadow and pituitary fossa is small and closed-in. Improved markedly on pituitary feeding.

complain. Concomitant with this headache, and due to the continual enlargement of the pituitary gland, growth is rapid. We then have a combination of symptoms, some due to the subinvolution of the thymus gland, such as genital underdevelopment, with pubic hair of invert type, together with a measure of fatigability and excessive growth, and added thereto frequent and periodic headache of migrainous type. The blood picture also slowly changes; the blood sugar content rising, the polymorphonuclears increasing and the carbon dioxide coefficient increasing.

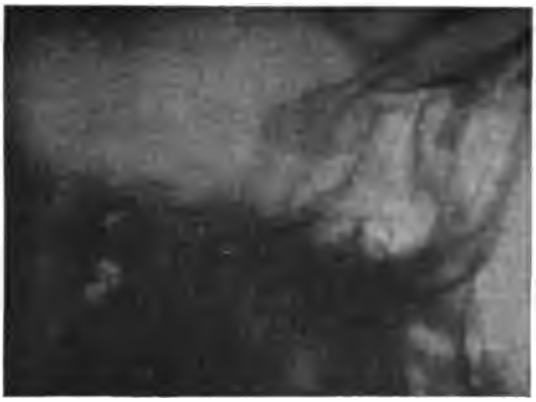

FIG. 24. RADIOGRAPH OF PATIENT IN FIG. 23. Small and closed-in sella turcica.

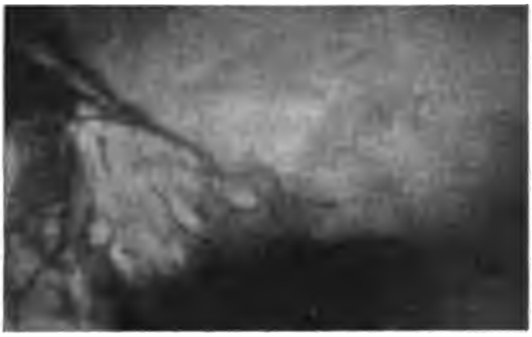

FIG. 26. *X*-Ray of very small and completely closed-in sella turcica in patient that never compensated for an early thymic condition; had all evidences of this state—late puberty, enuresis, intense headaches, great fatigability. Developed unmorally and committed sexual excesses. Pathological liar. Dipsomaniac also. On pituitary feeding, lost headaches completely and there is a marked return to a sense of responsibility and morality. Age thirty-two years.

Finally, when the pituitary has acquired all the margin of space it needs, due to the increasing size of the sella, the headaches cease spontaneously, and growth ceases. The blood pressure and blood picture become normal and the individual has emerged from his status situation to one of compensation quite sufficient for most needs. But in this process he has grown excessively in height, or per-

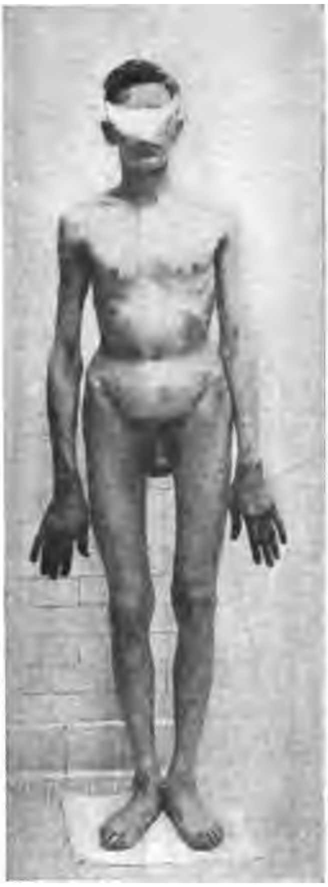

FIG. 25. FULLY COMPENSATED CASE EVOLVED FROM A BEGINNING STATUS THYMICO-LYMPHATICUS. Age thirty-two years. Gives complete history of early thymic state. Headaches persisted in adolescence with great weakness, concomitant with rapid growth. Now a few inches over 6 feet in height. Shows feminine pubic hair, feminine waist; never shaved; disproportion between thorax and legs in length. Large hands. *X*-Ray shows an enlargement and erosion of the sella. On pituitary feeding, with some suprarenal extract, has finally reached a stage where he can work. Supports a family of wife and two children; was invalided for eight years.

haps has even acquired some acromegalic features. Such acromegaly needs no treatment for it is not in itself a disease process, but the accentuation of a physiological one. And it terminates spontaneously. Frequently we see in the hospital patients who have been admitted for quite other ailments, but who show such evidences of earlier growth or acromegaly. Going over their earlier history, it is common to find the story of rapid growth, headaches, and great fatigability lasting through many years of their adolescent life and finally ceasing together. They are of the thymus-adrenal-pituitary syndrome that has just been described.

It can now be readily understood that the transmission of an endocrine defect need not take on the external characteristics of the parental trait, for each individual will compensate for the inherited defect in his own way, if he compensates at all. And so a parental trait which manifests itself in the thyroid domain, may be metamorphosed into one with pituitary or adrenal manifestations. It is of extreme interest to examine successive generations of patently endocrinopathic parents and see what a variety of pictures of a disturbed internal glandular system their compensations determine. Such a table, in conclusion, I have introduced here. From it I think we may deduce the fact that in just as diverse ways as the individual helps himself over his endocrine difficulties, so diverse in different individuals, apparently suffering from the same endocrinopathy just so varied must be our therapeutic attacks.

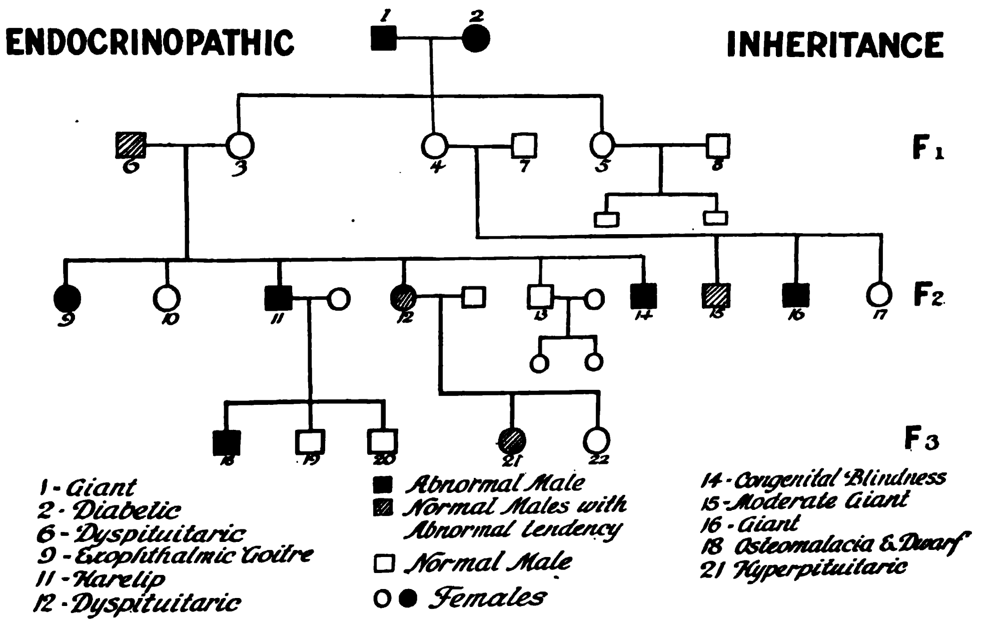

FIG. 27. CHART SHOWING THE ENDOCRINOPATHIC DEVIATIONS THROUGH SUCCESSIVE GENERATIONS OF AN ORIGINAL BI-PARENTAL DISTURBANCE. The original ancestors were afflicted respectively with giantism and diabetes.

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MENTAL HYGIENE AND EDUCATION*

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The subject of mental hygiene has not received the attention it deserves at the hands of educators and teachers, largely because of the prevailing notion, even among educated people, that the hygiene of mind is concerned with those whose minds have become diseased, or who have defective minds, that is to say, with the insane and the feebleminded. That those who have normal minds might also be benefited by mental hygiene, is an idea that seems difficult to grasp. Some explanation for this is to be found in the history of the mental hygiene movement itself. It is perfectly understandable how this movement should have received its strongest impetus in this country and elsewhere as a result of the facts and needs revealed in connection with the management of the insane and feeble-minded. It was the study of the diseased brain which gave us the most hopeful clues to an understanding of the normal brain structure and function—and in the disorganized, queer and perverse conduct of the insane there lies many a valuable suggestion for a practical hygiene of the mind. Thus it is that nature's own experiments in disease have been found to be of more practical help for the understanding and control of human conduct than were the laboratory experimentations of a psychology that concerned itself chiefly with a refined and precise study of the sense organs on the one hand, and on the other, with introspective speculations of an abstract or metaphysical kind concerning the nature of man. The principle is becoming more and more widely accepted that those are the best guardians of the body and mind in health who have some understanding of the nature of disease. If it be therefore suggested that an acquaintance with the principles of mental hygiene might find quite a natural abode in the equipment of the teacher, the explanation for this lies in the assumption that the teacher is naturally concerned with the mental health of her pupils. For no intelligent person would presume to dissent from the view that the process of education is intended to direct and shape the activities of living beings, that it is to assist human beings in the management of the business of living—of adaptation to the demands of the world about them.

And yet this view is being challenged in a very real and telling manner by the astounding fact that so many hundreds and thousands and hundreds of thousands of our citizens have to be re-educated later on in sanatoria, hospitals for the insane, jails and prisons. The management of the social problems created and kept alive by this huge army of failures in the business of living imposes a staggering burden upon the nation. It is an army responsible for more misery and sorrow than is even a world war, for death and bereavement are preferable to the lives that many of these unfortunates are obliged to live, and it carries with it a very serious menace to the future of the human race.

It is not my intention to exaggerate matters. There is no need for exaggeration. All we have to do is to open our eyes to the real situation in order to appreciate the enormity and seriousness of the problems of these socially maladjusted, who have failed to learn the business of living, the great majority of whom were former wards of the schools and colleges. We are receiving annually into our prisons, reformatories and jails some five hundred thousand men, women and children. We are constantly besieged by new demands for housing facilities for the feeble-minded and epileptic, and if the rate of increase in the number of those suffering from one form of mental disease—known as dementia praecox—should continue in the future as it has in the past, the State of New York, for instance, will require a new large hospital for the housing of this class alone every three years.

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*Lecture given at the Summer School, University of North Carolina, June, 1929.

The growing interest in these matters and the keener appreciation of our own share, as fathers, mothers, and teachers, as citizens and legislators in the maintenance of this sad situation, is based partly upon a growing freedom from the paralyzing influences of dogmas of predestination eugenic and other, and partly upon the real promise of relief which lies in an intelligent and spirited endeavor to bring principles of training and education more closely in accord with the demands of life. Of what use is the assiduous sharpening and polishing of the instrument of human adaptation, the mind, if no guidance is given in its use for the prime necessity of making one's way through life? It is being recognized more and more widely that the process of education should prepare students for life and not merely convert them into receptacles for the storing up of miscellaneous forms of information. To quote Dr. C. Macfie Campbell: "The breakdowns in life, the nervous invalidism, embitterment and depression, misinterpretation and accusation, ill-balanced enthusiasms and fads of religious, philanthropic, intellectual or æsthetic nature, family disharmonies, and futile day dreaming, do not arise because the individual has studied Homer and Euclid instead of Spanish and the Stock Exchange reports, or because he has followed the methods of the kindergarten instead of expressing his individuality in a Montessori atmosphere. The failures are more influenced by the fact that in the developing individual less attention is paid to the personal problems of the child than to the accumulation of information, less attention to the instincts, emotions and interests, the mental conflicts than to memorizing and facile repetition; yet during this period habits are formed which are to be of crucial importance for the happiness and efficiency of the adult."

"If education is to fulfill its task of preparing the individual for the business of life —if education is to continue to be defined as the intelligent direction of human activities, education must rid itself of the dangerous sophistry that tends to cast suspicion upon any system of education reflecting either utility of purpose or immediate practicabilty of application. The actual demands of life are full of these practical needs, and the value of ideals is commensurate with their practical usefulness." (Paton.)

My justification, therefore, in speaking to you on this subject rests upon the general belief that education has something to do with preparation for life, and upon a personal conviction gained after a great deal of experience in the management and treatment of human failures, that no educator can successfully fulfill this important phase of his task unless he is concerned with the total individual, with his reactions as a social being, called upon to adjust and adapt continuously to the demands of life, and not merely with his receptacles of information.

In presenting the subject it will be my aim, first, to define for you the mental hygiene movement—its aims, tendencies, and history of evolution; second, to bring to your attention some of the thoughts expressed by educators and mental hygienists on the relations of mental hygiene to education, with the view of making available for you such practical suggestions as these expressions may contain for teachers, and third, to outline the place of the institution for higher learning in the mental hygiene movement.

As to our first aim, the mental hygiene movement had its inception about a dozen years ago as a result of the efforts of Mr. Clifford Beers, who had himself suffered an attack of mental disturbance and who upon recovering from his illness published a book —"The Mind That Found Itself." This book contained not only extremely interesting and useful information concerning some of the phenomena of a diseased mind, but it served also to focus attention upon the intense suffering of the insane, and upon the unpardonable neglect of society to give the insane the attention that is due them.

The book came at a time when social forces and movements for human welfare reflected a growing consciousness of human needs, not alone in the direction of conserving human life and the prolongation of the span between the cradle and the grave, but also in the direction of deepening and broadening the meaning of life. Interest broadened from mere questions of length of life to questions of content and character. The innate human craving for this deeper and broader and finer expression of life naturally was there long before it became crystallized in social movements in that direction, but the birth of the mental hygiene movement helped to energize and organize the vague gropings in the direction of improving the content and direction of human life, since it had for its chief interest the focusing of attention upon the mind, the organ of human adaptation. It was also able to bring forth a prodigious volume of proof in substantiation of its contention that lack of attention to these finer needs of man is responsible for much of that disorganized and misdirected human energy which finds its outlet in destructive channels, leaving behind as it goes along the various social problems with which modern society has to cope. It challenged a reshaping of aims and helped to concentrate attention more definitely upon the real object of human concern in all its activities—mankind.

May I be permitted to quote from Dr. Barker's excellent statement of the relation of mental hygiene to the science which is concerned with the collective activities of man—Sociology? He says, "A glance at the data of this science and the truths that have been discovered regarding the structure, the functions and the genesis of society will convince anyone, I think, of the importance of a consideration of the relationship of sociology to mental hygiene. The Sociologist, as W. " pointed out, inquires into the conditions of human nature that induce man to enter into a state of society, into the results that social relations determine in modifying his feelings, his interests, his desires and his conduct, into the influences that tend to ever closer association and cooperation for ever multiplying purposes; into these purposes themselves and the machinery invented for serving them; into the many social structures that develop as a result of associated activity and into their functions; into the differences in these structures and functions in different walks of society and into the effects of each of these upon the intellect, the character and the conduct of man."

The social forces are described as consisting of the feelings and desires of the social units. These impelling motives include all the appetites that are concerned with individual preservation and with race continuance, and in addition all the desires, æsthetic, social, intellectual, that have to do with race elevation. The social forces are accordingly psychic phenomena; they belong to the feeling and striving systems of the mind and are to be looked upon as the dynamic agent that produces movement and change in the social domain, whereas intellect, or the knowing system of the mind, is to be looked upon as the directive agent that guides the dynamic agent into safe and effective courses of behavior. As social forces continue to conflict and collide, states of comparative equilibrium result and social structures like the family, the clan, the tribe, the state and the church arise along with social institutions like language, marriage, government, customs, ethical and conventional codes, religion, art, literature and science. In studying the functions of all these structures and institutions, and in observing how they are gradually modified and transformed into different and more efficient structures, the sociologist is impressed with the view that it is effort to remove obstacles in the way of satisfaction of desires that slowly produces the changes in social structures and institutions and is the cause of all social progress. Social control must then have as its first prerequisite a clear understanding of the feelings, the emotions, the desires of men—if these are to be consciously directed into desired channels—in order that society may benefit by them. It is the task of psychologists and sociologists to use the intellect to discover the laws to which human desires conform, and statesmen have the task of using the intellect to invent means of inducing men to act for the good of society—of "making it advantageous to the individual to do that which is socially beneficial."

It is inconceivable how this task can be achieved without a knowledge of man's emotions, desires, and tendencies, and yet it is this phase of men's nature—the dynamic force of progress, individual and social, which is most often neglected in the training of the child.

The task of the mental hygiene movement has been defined as "a continuous effort directed toward conserving and improving the minds of the people, a systematic attempt to secure human brains so naturally endowed and so nurtured that people will feel better, think better and act better than they do now."

It must be clear to you as teachers—as society's representatives in the promotion and the cultivation of means for the intelligent direction of human activities—what your place might be in this enterprise of getting people to feel better, think better and act better.

How modern education actually employs its opportunity in this direction, cannot be readily estimated. But the challenge contained in the phenomenon that literally hundreds of thousands of our citizens have to be re-educated in institutions which society has been called upon to devise for the care and management of human wreckage, deserves your serious attention. I am not an educator and it is not for me to enter into a detailed discussion of the educational process; but I have had innumerable instances in my dealings with the socially maladjusted where I have had the strong feeling that had the teacher had a better appreciation of and a better opportunity to concern herself with the driving forces in human nature, instead of having restricted her interest merely to the intellectual side of her pupils, much damage in the lives of these individuals would have been prevented.

Mr. Cook's statement in his book called "The Play Way," has found its justification in the histories of many a human failure. He says: "When will schoolmasters realize that because of their iniquitous preoccupation with their 'subjects' more than nine-tenths of the growth of a boy's experience is going on without any influence from them? When will they realize that a boy is somehow, or anyhow, adjusting himself with life quite apart from all their school teaching? Because of their lack of sympathy and contact with a boy's real interests he is all the time out of their reach. Let any school master honestly consider the boys he is influencing, and he will find them to be those whose interests he shares, those in whose confidence he is, and these will not necessarily be the boys who are any good in his subject. A master's educational influence often has very little to do with the subject matter of his teaching, and sometimes none whatever."

To sum up then, the aims of the mental hygiene movement might be said to be the encouragement and promotion of research into the nature of man and into the underlying facts of human conduct, and the dissemination of this knowledge for purposes not alone of ameliorating the lot of the insane and the feeble-minded, but for bringing about better feeling, better thinking and better acting in all people. The success of this enterprise depends very largely upon the wisdom and attitude of those who come in contact with the individual in the first decade of life, the parent and the teacher in the primary grades.

Coming to the second point in our paper, I can do no better than quote to you what one of the wisest educators of the age, Prof. Burnham of Clark University, has to say on the subject. In a very instructive and delightful paper entitled "Success and Failure as

Conditions of Mental Health" he discusses some of the weak points in our educational system. He devotes himself to a discussion of those educational fallacies which expose the child to continuous failure and deprive him of the stimulating influence of success. To appreciate fully what this habituation in failure really means one must be acquainted with the human wrecks who owe their failure to a crippling sense of personal inadequacy or inferiority which is frequently engendered by this early deprivation of the stimulus of success and by the exposure to failure. To quote Dr. Burnham, "The child from infancy has been matching motor images with reality over and over again and achieving a remarkable success in motor development, and has also been achieving social success in many ways; then he is put in an environment where success is very doubtful and many children instead of achieving success experience for quite a time the humiliation of failure." For many children in the school there is little opportunity for success. Failure in certain lines of effort is desirable but failure in everything is disastrous. Among school conditions and practices that tend to produce failure are the following:

Without examination, all of the children of the chronological age of six, the developed and the undeveloped, the sick and the well, the feeble-minded and the normal, the defective and the sound are received into the school. As a result the teachers are handicapped, many pupils are given work for which they are not fitted, either physically or mentally, and a large percentage of retardation and failure is insured from the outset.

The devotion of teachers to the subjects they teach as the thing of prime importance often means the inevitable neglect of the object of instruction, the boys and the girls to be taught. This tendency to consider first of all the subject and content of culture, and to put the object of culture in a secondary place is found everywhere.

Again, the school minimizes the opportunities for success by the large heterogeneous groups organized into classes. In a large class it is almost inevitable that the teacher should be handicapped and forced to give attention chiefly to the dull and delinquent pupils, or to those especially bright, or to divide her attention in a loose and inefficient manner over the whole group in a futile attempt to do justice to all. The school does not differentiate properly for exceptional children. Unusual children and strangely enough those that are specially bright, often prove failures in the school or are retarded. Dr. Terman has specially emphasized the fact that the bright are often retarded; and as everybody knows, the children interested in doing rather than in books prove failures. To the ordinary school, often enough, a conventional failure is more desirable than a troublesome success.

Most schools do not care properly for defective children. Among the pathetic tragedies of childhood are the cases of those who never can achieve success because of defect —the child with defective vision who cannot see the blackboard, the deaf child who cannot hear the teacher, the child tormented with headache or toothache, the child whose brain nutrition is reduced by nasal obstructions, the sensitive child, the misunderstood child, and the whole list of nervous defectives. The prevalent custom of offering prizes for the best work in different subjects, and the like, gives the opportunity for two or three to succeed, but all the rest must necessarily fail. It would be far better to offer a reward for all who succeed in reaching a certain standard of excellence, just as in some of the best summer camps, instead of a prize for the best, every boy who can swim a certain distance, or does a certain amount of work receives a badge of honor.

The fetich of symmetrical development causes many failures. A false pedagogy has exalted the value of an all-round symmetrical training and of the advantage of doing what children are not well fitted to do, and in which they are bound to fail.

How common this practice is, everybody knows. In illustration of this point, Prof.

Burnham tells a very charming story which I will quote.

"In antediluvian times while the kingdom was being differentiated into swimmers, climbers, runners and fliers, there was a school for the development of the animals. The theory of the school was that the best animals should be able to do one thing as well as another.

"If an animal had short legs and good wings, attention should be directed to running, so as to even up the qualities as far as possible. So the duck was kept waddling instead of swimming. The pelican was kept wagging his short wings in the attempt to fly. The eagle was made to run, and allowed to fly only for recreation.

"All this in the name of education. Nature was not to be trusted, for individuals should be symmetrically developed and similar, for their own welfare as well as for the welfare of the community.

"The animals that would not submit to such training, but persisted in developing the best gifts they had, were dishonored and humiliated in many ways. They were stigmatized as being narrow-minded and specialists, and special difficulties were placed in their way when they attempted to ignore the theory of education recognized in the school.

"No one was allowed to graduate from the school unless he could climb, swim, run and fly at certain prescribed rates, so it happened that the time wasted by the duck in the attempt to run had so hindered him from swimming that his swimming muscles had atrophied and so he was hardly able to swim at all; and in addition he had been scolded, punished, and ill-treated in many ways so as to make his life a burden. He left school humiliated, and the ornithorhynchus could best him both running and swimming. Indeed, the latter was awarded a prize in two departments.

"The eagle could make no headway in climbing to the top of a tree, and although he showed he could get there just the same, the performance was counted a demerit, since it had not been done in the prescribed way.

"An abnormal eel with large pectoral fins proved he could run, swim, climb trees and fly a little; he was made valedictorian."

From this prehistoric attempt at ideal education down to the present many schools have put a premium upon failure. And from this continued effort to do work that cannot be done well, pupils are apt to acquire a slovenly attitude, and like the young horse given too heavy a load at first, they acquire the habit of lying down under a difficult task.

The relation of this chronic discouragement to the habitual dodging of issues in real life, and to those evasions, faulty compromises and gradual withdrawal from reality which characterize the dementia praecox patient is a close one, and these faulty biologic habits of reaction are frequently formed very early in life.

But discouragement and failure may lead to other forms of reaction than those characteristic of individuals who are overburdened with a sense of inferiority and who have succumbed to it. The individual may react to this feeling of inadequacy by the development as an over compensation of an undue haughtiness, aggressiveness and egotism. Many a bully in school reflects merely an attempt at compensation for an innate sense of inferiority or defect. If the school and home do not provide the means for experiencing the satisfaction and thrill of success in socially desirable fields, the boys and girls are apt to find it in mischief and destructiveness. The tendency to seek for the cause of the maladjustment or difficulty solely in the boy, on the naïve assumption that nothing in the school or the home can possibly have anything to do with the boy's mischief, is responsible for much of our ignorance concerning these matters. Truancy, as Campbell puts it, may be, of course, the reaction of an unintelligent child to an intelligent school system; but it may also be the reaction of a rather exceptionally intelligent child to an unintelligent régime. Disciplinary difficulties may have their roots in the manner in which the discipline is exercised. To want a child to do a certain thing merely because the parent or teacher says so, may not be a sufficiently attractive reason for the child to carry out the command. On the contrary there is just enough temptation in the situation to make the child do just the opposite. The same is true of the effects of punishment and reward. Unless the child is made to understand clearly the aims and objects of the reward or punishment—unless the association between these and the specific act of the child is made absolutely clear to him, these measures of control are worse than useless. Unfortunately parents and teachers employ these methods often enough more for personal satisfaction than for the purpose of instilling in the child a lesson in conduct.

Moreover, the manner of exercising parental or pedagogic authority may affect the child's attitude toward all forms of authority and may make of him, as Campbell says, a truculent and unwilling worker, defiant of all authority, a fomenter of discord and a nuisance to himself and others.

The avoidance of serious errors in the conditioning of the pupil's conduct and attitude as a social unit will always depend upon the ability of the teacher to see in her pupil not merely an assemblage of organs plus a receptacle for school information, but an individual with a complete mechanism of highly organized instincts, emotions, strivings and attitudes which are the real driving forces in conduct.

Unfortunately in the training of the teacher very little attention is being given to the subject of mental hygiene. Until a radical improvement takes place in college and normal school curricula our teachers will continue to be hampered and inefficient in those matters which concern the child most vitally. Indications of a greater recognition of the importance of instruction in mental hygiene are already at hand. There is an ever increasing demand on the part of the teachers themselves for specific information in this field. It remains to be seen how soon educational authorities will provide means for the achievement of this. One cannot enter here into a detailed discussion of what this education is to consist of but it should be clear that we have not in mind the traditional categorical and descriptive presentation of mental disease and defect.

What is needed for purposes of a workable hygiene of mind is a discusion of mind in the making, in the "becoming" in the sense of Bergson. Much dependable knowledge is available concerning the basic structure and the fundamental mechanisms of the human nervous system to make possible a truly helpful consideration of the driving forces of human conduct and of the conditioning influences which shape personality. Such a presentation would involve the discussion of the nature, varieties, and modes of expression of the instincts and emotions, of the organization of these into the higher and more complex systems of the mind—the sentiments which have so much to do with determining man's conduct as a social being, of the conflicting issues which constantly arise between the blind, innate animal cravings of men and the directive and controlling tendencies of the sentiments in the individual's endeavor to adapt himself socially, of the compromises, evasions, and vicarious substitutions or more or less adequate solutions to which these conflicts lead. It is this type of discussion of the moving, dynamic issues in human behavior and in a more descriptive psychiatry which deals with static entities in mental disease and defect that is needed for a furtherance of the hygiene of mind. The present curricula of colleges and even medical schools make little or no provision for this kind of presentation, so that if the aims expressed in this paper are to be fulfilled there is needed an emancipation of psychiatry from the traditional narrow pathologic view point on the one hand and on the other a broadening of psychology to include those aspects of the workings of the human mind which have come to light as a result of the efforts of the newer dynamic and analytic psychology.

BROWN-SEQUARD SYNDROME PRODUCED BY AN OLD CERVICAL TUBERCULOUS SPONDYLITIS*

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This case is presented before the conference not only because of its clinically interesting features, but also because it illustrates the importance of following a detailed outline, such as is now being employed in this clinic, whenever a patient is examined neurologically. Only after a thorough physical, neurological, cytological, and roentgenological examination of this case was its true nature detected, and the examiners enabled to avoid repeating the mistake which occurred three years ago of regarding it as of secondary importance.

CASE No. 27161. G. R., male, Russian Hebrew, twenty-eight years of age, married, eight years in the United States, came to the Neurological Clinic by transfer from the Medical Clinic on January 21, 1920, complaining of stiffness in the right hand and in the right foot which, he claimed, had followed a blow on the base of his right index finger while nailing down the cover of a trunk three weeks before.

Past History.—He knew nothing of his infancy or childhood. He had attended a Hebrew school from eight to twelve years of age and claimed to have been a good student. At fourteen years of age he had an abscess in the right side of the neck which opened spontaneously but which continued to discharge pus for at least a year. During this time he was confined to bed, being paralyzed on the right side and having difficulty with his speech. No other details of this illness could be elicited. When twenty-five years of age the patient contracted gonorrhea and was treated for a period of six months. He married at the age of twenty-six and his only child, a girl of nine months, is in good physical condition. For the last year he has been practicing coitus interruptus.

Family History.—This, as far as the patient knows, is negative for three generations.

The patient was very evasive while giving the anemnesis. He denied ever having been to this or any other clinic before. He was dull, and appeared to have considerable difficulty in grasping the significance of the questions of the examiner although the language used was the patient's native tongue.

PHYSICAL EXAMINATION

VOLUNTARY MOTOR SYSTEM.

Abnormal Attitudes and Deformities.—The head was held rigidly erect. There was a bulging in the left side of the neck and in the left occipital region.

Gait.—In walking the patient revealed a definite spasticity in the right lower extremity.

Coordination.—The equilibratory coordination tests were normal. There was no Romberg and the patient was able to stand properly on either foot. The non-equilibratory tests, as shown by the finger to finger, finger to thumb, and toe to object tests, revealed a normal response in the left, with definite ataxia in the right side. The right finger showed definite past pointing to the left. Heel to knee test showed a normal response on the left side, and ataxia on the right. The result of the tests was the same whether performed with the eyes open or closed. Adiadochokinesis was present in the right and doubtful in the left. Dysmetria was present in the right and doubtful in the left. There was more of a rebound in the right than in the left.

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*Presented before the Conference of the Neurological Department of Columbia University on February 4, 1920.

Skilled Acts.—These were performed rather awkwardly. The speech was thick and rather slurring. Test phrases showed no formal speech disorder.

Abnormal Involuntary Movements.—These were seen in the form of fibrillary twitchings in the right shoulder and in the left abdominal muscles.

Reflexes:

Deep

	Left	*Right*
Jaw	0	0
Pectoral	2	2
Biceps	2	3
Triceps	3	3
Radial	3	3
Ulnar	3	3
Wrist	3	3
Suprapatellar	3	4
Patellar	3	3
Patello-adductor	R to L 1	L to R 1
Tibio-adductor (periosteal)	2	2
Tibio-adductor	R to L 1	L to R 1
Cuboido-digital	0	0
Achilles	4	5
Ankle clonus	transient	permanent

Superficial

	Left	*Right*
Ciliospinal	0	0
Supra-umbilical	2	2
Supra-pubic	2	2
Upper lateral abdominal	2	2
Lower lateral abdominal	2	2
Cremasteric	1	1
Babinski (toe)	0	present
" (fanning)	0	present?
Chaddock	0	present?
Oppenheim	0	present?
Gordon	0	0
Schäfer	0	0

O = absent, 1 = slight, 2 = active, 4 = with transient clonus, 5 = with permanent clonus.

Muscle Strength.—The patient was unable to bend his head forward and backward and was definitely limited in moving it from side to side. There was slight atrophy of the right biceps muscle. Otherwise the musculature was of normal consistency and its tone normal throughout.

Abnormal Associated Movements.—Pronation signs of Strümpell were present in the right, and Hoffman's sign was present in both right and left.

Nerve Status.—This was normal.

General Sensory.—This status was difficult to elicit because of poor cooperation on the part of the patient. He showed, however, definite diminution in the pain and temperature senses on the left side, including the left side of the face, and a possible diminution in the muscle tendon and vibratory senses on the right side, particularly in the lower extremity and especially in the toes. At times, there seemed to be a slight right astereognosis.

Cranial Nerves.—The olfactory nerves were normal.

Each eye showed perfect vision and the fundi presented nothing abnormal. The pupils were unequal, the right being larger than the left; they reacted promptly to light and to accommodation. The right palpebral fissure was somewhat less than the left. There was no nystagmus and the movements of the eyes were not limited in any direction.

The acoustic nerve and ear showed nothing abnormal.

The trigeminus nerve was intact except for slightly impaired corneal reflexes. The mouth showed an irregular setting of the teeth and congested tonsils.

The taste was normal.

The ninth, tenth and eleventh cranial nerves showed nothing abnormal. The twelfth showed defect in the rather awkward movement of the tongue and in the thick articulation.

Mental Status.—This will be discussed in further detail later on.

Cranial Morphology.—Nothing abnormal was found.

Systemic.—General systemic examination showed palpable thyroid, large tonsils, and enlarged inguinal nodes. The heart was of normal size, was regular in rate and rhythm and there were no murmurs. Blood pressure was 125/80. Lungs were negative. The gastrointestinal tract presented nothing abnormal. There was a large left inguinal hernia.

LABORATORY TESTS

Wassermann of the blood and of the spinal fluid also was negative. There were no cells in the spinal fluid and the globulin was negative.

Roentgenological examination of the lateral and antero-posterior cervical region showed a scoliosis of the upper cervical spine due to an old healed Pott's disease in the bodies of the second and third cervical vertebrae. (Figs. 1 and 2.)

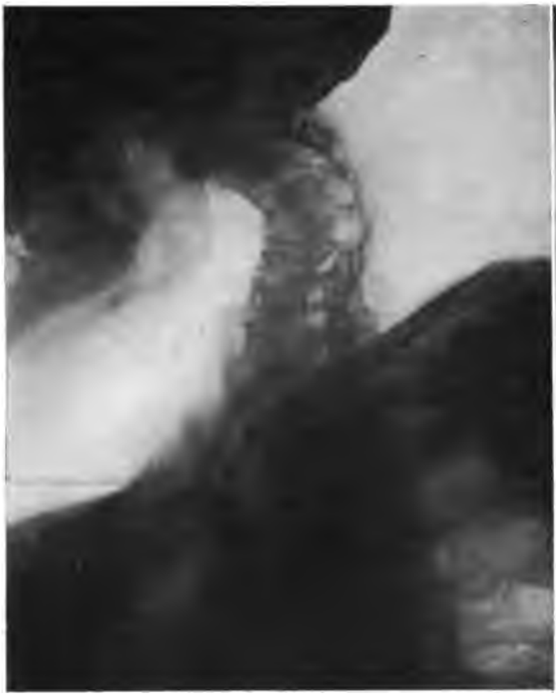

Fig. 1. Lateral View of Cervical Spine.

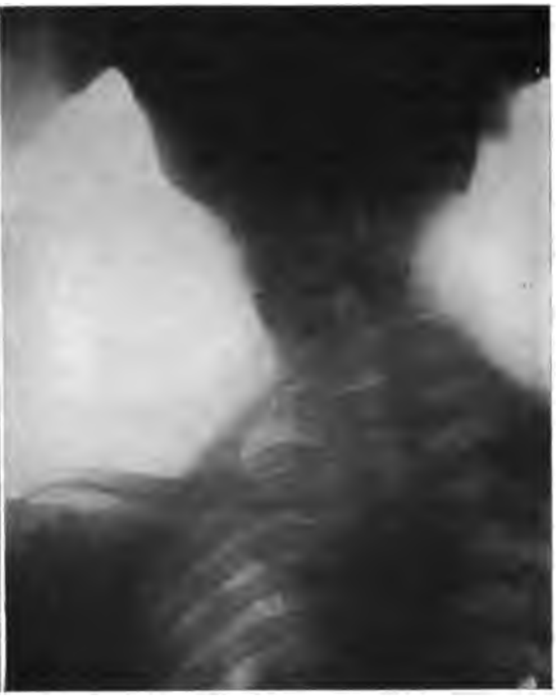

Fig. 2. Ventral View of Cervical Spine.

To refer back to the mental status, in this regard the patient presented rather interestting phenomena. His general attitude and manner were those of an individual constitutionally rather than mentally defective. His failure to grasp the significance of the examiner's questions was not due to an inherent mental defect, but to an unusual suspiciousness which seemed to dominate his entire conduct. He was very slow in answering, weighing each reply carefully and with most apparent shrewdness. His attitude towards life was that of an individual to whom the world was indebted because of his mere presence in it. Winning his confidence to a certain extent brought out the confession that his sole aim in visiting the Clinic was to gain the interest of the social service worker to help him look after his family as he did not feel well enough to work.

Emotionally he was rather unstable; his mood was that of depression alternating with definite exaltation. It was hardly adequate to the stimulus eliciting it. There were no delusions or hallucinations. Both for remote and recent events the memory was good, and the grasp on current events as well as general school knowledge was very good. The patient, however, had no insight into his condition.

His attitude towards the examiner and towards conditions in general is well illustrated by the fact that his physical condition indicated an old standing disease which must have given him trouble on some previous occasion and which would lead one to believe that he must have visited some clinic or dispensary, and yet he persisted in denying that he had ever done so. He was, however, recognized by a worker in this Clinic as having been here for treatment in 1917. When this was called to his attention he repeatedly refused to acknowledge the fact, and only admitted it when confronted by the record of his old history. The history was recorded as follows:

"CASE No. 0650. Feb. 7, 1917, age twenty-three, single, male, complained of pains in the gums and headache which had come on four days ago. The pain came on suddenly in the gums and the right side of the head and face and was of a constant nature. He also complained of dyspnea and of frequent diurnal urination. Physically nothing abnormal was found except filthy teeth, fetor oris, caries and calcareous deposits on teeth, and a slight tremor of the hands. Reflexes equal and normal. Sensory examination normal. Diagnosis: torticollis."

The outline followed at the time of this earlier examination was definitely inadequate and, while satisfactory for grosser neurological cases, entirely useless for such subtle cases as this one.

SUMMARY

This case, a man twenty-eight years of age, married, a Russian Hebrew, constitutionally inferior, who came to this clinic in 1917 complaining of pain in the gums and general headache and who then, according to the record, showed nothing abnormal except tremor of the hands, filthy teeth, fetor oris, and caries and calcareous deposits on the teeth; was diagnosed as torticollis; returned to the clinic in 1920 complaining of stiffness in the right hand and foot; on physical examination showed definite twitching in the right shoulder muscles, definite right-sided non-equilibratory ataxia, right-sided Babinski, and transitory supra-patellar clonus, right abnormal associated movements, diminution in temperature and pain senses in the left side except the face, and diminished muscle tendon sense in the right lower extremity especially the toes; negative blood and spinal Wassermann reaction; and, roentgenologically, scoliosis of the upper cervical spine due to an old healed Pott's in the second and third cervical vertebrae. The lesion of the cord was in the cervical region, mostly right-sided and mostly lateral, the tracts involved being the right spino-cerebellar, the right spino-thalamic, the right pyramidal, and the right columns of Goll and Burdach, causing the above symptoms.

THE LEONARD PRIZE FOR RESEARCH

THE AMERICAN ROENTGEN RAY SOCIETY WILL AWARD $1000.00 to the author of the best piece of original research in the field of the *x*-ray, radium or radio-activity. The competition is open to any one living in the United States or its possessions, in Canada, Mexico, Central or South America, Cuba or other islands of the Western Hemisphere. The research matter must be submitted in literary form in the English language not later than July 1st, 1921, and must never have been published. Each paper must be signed by motto and accompanied by a sealed envelope containing motto and name, so that the identity of the author may be disclosed after the award has been made. In case a demonstration of an invention or method is necessary, the identity of the author may have to be known before the award is decided.

The field of research includes discovery, invention, improvement of method, or investigations to prove or disprove any theory or problem, whether old or new, which has a direct bearing upon the use of *x*-rays, radium or other radio-active substances.

This competition is not open to any research involving a patent or copyright, and the submission of any research work for this prize carries with it the understanding that the subject matter will remain open to free use for the general good.

The committee reserves the right to withhold the award if in its judgment no piece of research offered possesses sufficient merit. It also reserves the right to divide the award if two papers are judged to be of equal merit. A certificate of honorable mention will be given to the author of meritorious research which has been submitted but which has not received the prize.

Papers, records or material are received in this competition only with the understanding and agreement that any work receiving award or honorable mention may be published in The American Journal of Roentgenology at the discretion of the research committee. All expenses of publication will be borne by the Journal. Papers not published will be returned to the authors.

The piece of original research receiving award must be presented before The American Roentgen Ray Society at its next Annual Meeting, September, 1921.

This prize is offered in an altruistic spirit for the promotion of useful research, with the approval of the National Research Council. It commemorates the name of a martyred member of the American Roentgen Ray Society, Dr. Charles Lester Leonard, who paid the supreme penalty for his pioneer research in the field of the *x*-ray.

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SPEECH, ITS DEVELOPMENT AND INTEGRATION WITH THE INTELLIGENCE

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PART I.

The subject of the development of speech in the human and its disturbances has always engaged the interest of medical men. Discussions of this subject have resulted in the alignment on opposite sides of those who adhere to the desire strictly to localize this function in more or less sharply circumscribed areas of the brain and those who have contended that speech is not so exactly localizable a function and that it depends on the working of the entire brain as a whole by the process of association and correlation. The most acrid controversy of recent medical history took place in France between the proponents of these two sides of the question and resulted in many bitter and sometimes personal criticisms. As is usual with controversies of this kind very little real benefit has accrued. Recent criticisms of the author's monograph on aphasia have shown that it is possible to begin the same sort of controversy in this country, providing that those who have the broadest conception of the speech functions are going to allow themselves to be distracted from a consideration of anything but the purely scientific side of this question.

The difficulty, it has seemed to me, has been that the two main elements in speech production are not always definitely separated. Articulate speech is the purely mechanical sound-producing function which depends on a preservation of the innervation of the muscles of the palate, larynx, lips and face, and quite as important, of the lungs and chest musculature including the diaphragm. Articulate speech therefore looked at in this way is a purely kinetic thing and has the same mechanics as any other skilled act. So too with writing and mimicry, which are the other motor forms of speech expression. Indeed one might with propriety include here the entire facial musculature and that of the trunk and extremities, for one can just as surely "speak" by assuming an attitude or by using various expressions of the face as one can by articulation.

The other, broader, element in the subject, is the matter of intelligent thought and its development. Speech is a function which belongs to gregarious animals and exists because of the necessity for conveying thought, which is simply another way of saying that only in this way can these animals communicate their desires and give expression to their needs and yearnings.

Speech is a learned function. Everyone must learn to speak. It develops apace with the intelligence. The expression of thought in speech, whether it be articulate or by the assumption of facial or other attitudes or by writing, is made possible through the integration of a series of symbols. These are mainly sound, visual, gesture (kinesthetic and proprioceptive), olfactory and tactile symbols. These symbols in order to be used in speech [illegible] a [illegible] meaning to the individual [illegible] of a group. All of the symbols are learned, however, and because the [illegible] training and environment varies [illegible] the method of learning these symbols [illegible] only be a variable process. The speech functions therefore develop hand in hand with all other forms of intellectual development and are an integral part of intelligence as a whole. To clarify this conception one need only turn to every day experience. The children who come to us ignorant and untaught from foreign shores, possessing so few of the symbols mentioned that speech in their own language is monosyllabic, or poor in its conception and abstractions, develop through the process of education and contact with a plenteous environment, a great number of the necessary symbols and soon are able to give expression to the thought thus developed in them in the most acceptable forms of our own language. The individual speaks best (using speech in the broader sense) who has learned most. How either of these processes can be definitely separated one from the other is something beyond our ability to understand. This function cannot therefore be localized in the brain in any such areas as have been diagrammatically taught us in the past. The projection centers in the motor sensory area of the cortex have an undoubtedly accurate localization, as have also the auditory, visual and olfactory centers. But speech in its broadest sense cannot be localized in any way comparable to these.

Articulate speech is a particularly skilled act and it results from a necessity which is inherent in all of us to express our desires and also our thoughts. Articulate speech, however, is only one means of expressing [illegible] thought in answer to our [illegible] The assump[illegible]

and this in turn depends on the number of his needs, urges, instincts and emotions and the necessity for giving expression to these. The broadening of intelligence and the development of intellect or mind is synthetically arrived at by the development of our reactions to the instincts and the emotions limited only by our environment and our powers of adaptation and assimilation. One notes that we have in this broad way also made it possible to include speech as a function possessed, possibly, by primates other than man. That there is sufficient basis for this possibility has been developed by various students of animal behavior. It has become a necessity for the investigation and understanding of many medical problems to look to comparative anatomy, physiology and psychology for a correct understanding of them. Attempts have long ago been made to understand human intelligence or the human mind by observing the psychological problems involved in their simplest state in animals. There is no good reason why our problem should not be attacked in the same way.

It does not seem merely a coincidence that of the various studies, the animals most frequently chosen were the apes and monkeys. It is a matter of common knowledge that these animals are the most intelligent in the animal kingdom excepting man. Of the various birds which have the power of imitating human speech or song, the parrot is not only the most excellent example, but combined with this he possesses as a necessary corollary the greatest intelligence. Green[1] has given numerous instances of extraordinary intelligence observed by him in both the parrots and parakeets. This author has made many observations of interest and quotes a number of incidents of their behavior which might be taken psychologically to mean that these birds not only have the ability to imitate but also are capable of associating articulate speech with symbols, recognition of persons and simple forms. These are simple but nevertheless unmistakable signs of thought processes. Many other incidents are quoted by Page[2] of the use of language by parrots, not only in a repetitive but also in a rudimentarily intelligent way. Time and again these authors observed what appeared to be unmistakable evidence that some of these birds were able to understand a very little of what they were saying and used language not merely as an imitative automatic cry without intelligence, but by means of association and experience gave some sign of understanding a few of the things they uttered, and of being able to give expression by utterance to certain emotional states. Green, who is a physician, on page 32 of his book notes that a solitary budgerigar parakeet will utter calls for a mate after a time and suffers from an acute depression which is termed pairing fever. If not relieved he will refuse food and die as a result of his depression. All of the parakeets described by Green show a distinct correlation between their intellectual powers and their ability to mimic sounds and speech. In other words, in direct proportion to the ability which they have to utter speech, does one find that these animals show other evidences of intelligence. Page, in the first 20 pages of his book, gives a description of the African gray and the Mexican double yellow parrots and definitely states that they are not only the best talking parrots but also the most easily taught and the most intelligent. He refers (p. 51) to the fact that they sing and whistle and speak better and possess a larger vocabulary the earlier they are taught, so that it seems quite definitely established in these animals that intelligence and the ability to articulate human speech go together.

A definite statement is made by these authors that the brains of parrots are proportionately much larger than those of any other animal. Page refers (pp. 180-182) to other talking birds, such as the magpie, raven, starling and crow and also to the talking myna, bluejay and jackdaw. He says that these animals have a less retentive memory than the parrot and also are undoubtedly less intelligent, and that their speech and even their power of imitating speech and sound are much more limited. Some of these animals are able to imitate sounds or even one or two words or phrases which however must be frequently repeated to them. All of these birds, Page says, can be taught a little speech if they are obtained early and trained diligently.

It is not my object to enter into a controversy of a psychological nature as to whether true thought in the human sense occurs in these animals, but it seems impossible to deny that they possess a certain degree of intelligence, and that the greater this degree the greater their ability to utter and to mimic human speech.

Regarding monkeys and apes and other infra-human primates, lessons similar to this can be learned. Unfortunately the only reliable observations on monkeys made by medical men were made with objects in view other than those which interest us. The attention of G. V. Hamilton[3] and Edward J. Kempf[4-5] was directed towards the social and sexual behavior of primates. However, in going through their work one finds undoubted evidences of an embryonic state of intelligence in these animals in the sense most often accepted by psychologists. Everyone knows that the powers of mimicry and imitation in primates is very highly developed. Kempf (p. 137) refers to the fact that monkeys will often protect their favorites from persecution by other monkeys even at the risk of severe punishment. Garner[6] refers to the fact that apes show solicitude when members of their family are ill and that they have a marked instinct of curiosity and are gregarious and that they appear to show gratitude and are able to express by means of speech states of danger and fear, and also are able to communicate their desires. Concerning Garner's observations more will be said later.

Hamilton states (p. 317) that the animals under his observation possessed the undoubted power to give expression to at least three different kinds of hunger or needs for individual satisfaction. These impelled the animals toward manifestations of sexual behavior and consisted of (1) hunger for sexual satisfaction, (2) hunger for escape from danger, and (3) hunger for access to an enemy. His conclusions and those of Kempf would mainly appeal to those interested in the psychology of the sexual instinct and are indicative of the low levels to which the minds of these animals have developed; nevertheless, undoubted signs of their ability to give expression to this low type of intelligence were found by these observers in the apes studied. A psychologist, S. J. Holmes[7] (p. 50) quotes Herbert Spencer as saying that "if the doctrine of evolution be true, an inevitable implication is that mind can be understood only by observing how mind is evolved." This author (p. 259) describes his experiments, which are simply a repetition of more extensive and more painstaking experiments on larger groups of monkeys observed by Kinnaman and Thorndike.[8] These psychologists by means of testing the ability to associate, as seen in the performance of definite experimental tests with glasses and cubes, locks, colors, forms and various types of trap apparatus, all of which were frequently repeated, attempted to gauge the intelligence of monkeys. It must be understood of course that these animals were transported to an environment which was entirely strange and that they were in captivity and either in complete solitude or in the company of only one or several of their own or other species of primates. Holmes' experiments (p. 259) led him to the belief that the mind of a monkey is developed by his instinct of curiosity and his reactions to the instinct of the acquisition of food and sexual satisfaction. As a result of numerous experiments with boxes, traps made of nailed boards, etc., all of which contained food the obtaining of which was the reward for the animal and acted as the stimulus which impelled him to follow out the experiments, Holmes concluded that his monkey was possessed of a quick perception, rapid judgment but no deliberate reasoning or abstract ideation. Mistakes were repeated, particularly if even mild difficulties were placed in the way of the accomplishment of the tests. His monkey seemed to learn simple things by chance success and by elimination of useless movements, which is also the description of the result of the successful experiments described by Thorndike. Holmes' monkey was only a small bonnet monkey, rather young, and as he says, the objection is always permissible that this monkey may have been an unusual individual possessing a personal trait, characterized by ineptitude. This experiment of Holmes had been performed many times before in a more thorough manner and with a greater number of monkeys by Kinnaman[8] and Thorndike[9] and many others. All of the experiments were similar to a certain degree. The animals were asked to solve the problem of getting food or freedom or sexual satisfaction (Hamilton) by overcoming obstacles such as escaping from intricately constructed wire cages, opening locks, choosing forms and colored candy, etc. As a result of his very careful experiments Kinnaman (pp. 200-213) believed that the monkey was able to recall by images, objects and persons not present to the senses. He also believed that his monkeys gave examples of the use of generic images such as Ribot called "intermediate abstraction." This is a higher form of mental process than the simple idea or notion or the simple memory of a particular sensory perception. In these examples Kinnaman felt that his monkeys were able to combine simple ideas into composites without the aid of language. This too is a fairly high degree of intelligence. He felt, however, that higher concepts and abstractions requiring the use of language were wholly wanting. He agreed with Ribot[10] that these animals possess the power of forming intermediate abstractions with merely "positions of the body or calls as signs of these." He believed that the faculty of abstracting even in these lowest forms, however, is the same process as the higher abstractions occurring in man, and that this development is dependent on the same process as that of intelligence and of language, and that "it exists in embryo in those primitive operations which are properly concerned with the concrete, i.e., perception and representation."

Without going too deeply into the psychologic intricacies of the problem, a careful review of the literature on animal behavior as it applies to the mental life of the infra-human primates seems to show that while these animals have not of course the power to form abstractions and concepts in the highest sense which we are in the habit of ascribing to human thinking, nevertheless they possess the very simplest beginnings of such powers and possibly even the faintest trace of language even though this may be limited to the few "calls" described by various authors. It does not seem necessary to follow out this matter any further; but perhaps one may do so without the danger of tingeing his subject with an element of unscientific romancing by calling attention to several publications by Garner.[11] His was the only work performed under conditions which even closely approximate the normal life and environment of these animals. His book (Apes and Monkeys: Their Speech and Language) is worthy of perusal, although one might wish that it had been written as a scientific contribution rather than as a popular narrative. This author (p. 19) was able to make out nine monophrastic sounds and one distinct phonetic sound in capuchin monkeys, all of which words, sounds, cries or calls, whichever one wishes to call them, seemed to be expressions of friendship, distress or fear. Garner agreed that monkeys and apes had no conception and that their thoughts were certainly not intricate; nevertheless that they were able to express their simple desires, he firmly believed. He believed also that monkeys and apes showed definite traces of sympathy and were possessed of a gregarious instinct and the instincts of curiosity and mimicry to a high degree. He believed that the speech of apes and monkeys as he heard it in their natural homes, the jungle in Africa, or in cages with experimental animals, was a voluntary articulate and deliberate use of sounds which they understood and which other animals of the same species replied to and understood. He believed that the higher apes, the gorilla and chimpanzee, had a higher type of speech than monkeys and that each race or species had peculiar calls of its own. He felt that the more gregarious the habits of any species the higher the type of speech it had (p. 23). Garner himself was convinced that monkeys and apes understood his imitation of their sounds and he also seemed convinced with Prof. Romanes[12] that these animals were able to enumerate and to distinguish numbers of objects and to find errors in the sum total up to the number of seven when incorrectly presented to them. He agreed with Kinnaman and Thorndike and others that the cebus was the most intelligent of the monkeys and that the gorilla next to man possessed the highest intelligence and therefore the most plentiful and most intricate form of articulate language. In the apes the rudiment of a paternal instinct is developed and there is a tendency to a patriarchal family life among them, and these apes are capable of showing solicitude when members of their family are ill, although this affective state is not maintained for a long time. The apes have a great number of words, 30 to 35 of which Gardner was able to understand and imitate. These words, calls or sounds, are expressive of gratitude, warning, desire for food or drink, or are expressions of discomfort, fear and greeting. He was convinced that chimpanzees also made negative signs with the head or were able to mimic the wavelike motion of the hand from the body as a sign of negation or refusal. He insisted that there was pitch and volume and tone to the voice of the apes in the production of their sounds, changes in which were regulated to suit the varying conditions under which they were used, and that apes understood the value of sounds as a medium of conveying thought. These observations have never been confirmed by a medical or non-medical psychologist and they have been viciously attacked and ridiculed both in the psychological and lay press.

It is with great hesitancy that one accepts all of these statements as facts; nevertheless all writers on this subject agree that apes and monkeys have several distinctive calls which correspond to definite moods or mental states expressive of certain instinctive needs. For our purpose this observation is sufficient.

No one can gainsay that the apes and monkeys are the most intelligent animals. It seems definitely established that they have a limited vocabulary of sounds which express certain desires and are used differently under various conditions. These sounds or calls are capable of interpretation by observers and by animals of the same species and are to all intents and purposes the rudiments of language. If one remembers that mimicry in man is also an integral part of the speech function, then certainly no one who has seen trained apes or monkeys perform on the vaudeville stage will deny that these animals possess this rudimentary and inferior form of speech and that they have it developed to a remarkable degree. Whether we believe with Romanes and Garner that animals are capable of processes of reasoning comparable to those of the human, or whether we believe with Thorndike who insists that animals cannot reason by analogy or abstractly, is entirely unimportant for our purpose. Careful observers like Kinnaman take a stand midway between these. He believes (p. 200) that the ruling out of reasoning by analogy cannot be entirely true of rhesus monkeys and that "in part their reasoning is like that of man." Nevertheless even if this were not entirely true, there undoubtedly exists in the monkey and the parrot an embryonic, a developmentally rudimentary trace of speech and intelligence which is later seen to best advantage and sometimes in its highest state of development, in the human. These animals probably have a dim consciousness of the resemblance of things and persons, and as Thorndike expresses it, "they learn by the elimination of useless movements and by chance success and by finding pleasure in the attaining of desires," but nevertheless they learn. That there is a limited emotional and mental life Kinnaman seems convinced. They indulge in movements and calls indicative of fright, anger, a desire for their fellows, curiosity and surprise. Their calls are particularly loud when they are separated from the company of their fellows. Their most active efforts are directed towards the attainment of sexual and hunger satisfaction. As a result of his experiments Kinnaman was convinced also that they made great progress and showed capability in choosing better methods for obtaining a gratification of these desires when obstacles were placed in their way by means of locks and other apparatus. He was convinced that they had to a considerable degree the power of learning by association and he felt that there were undoubted evidences also of learning by imitation, one example of which was of very high order (p. 211). He was also convinced that they liked and preferred and made known their preference for certain colors, yellow, orange and green being preferred to red, purple, blue and gray. He makes the statement that "human beings have not greatly surpassed them in some respects when dealing with the apparatus here used," and one must remember in considering his conclusions that he used rhesus monkeys which are not so intelligent as the cebus and certainly not so intelligent as the chimpanzee and the higher apes.

Going higher in the scale of animals we now come to the study of speech and its intelligent expression as articulate language in children. This whole matter may be summed up in the words of A. H. Ferrard [13] who says that "it is not until the baby begins to talk that one regards it as an intelligent unit . . . for in these early essays at talking a great deal of light is thrown upon the manner in which we all have learned to think and communicate our thoughts to others." Briefly, the first cries of the child are comparable to the calls of the monkey; they merely express desires and discomfort, and children repeat them by learning that these cries bring relief. As the child learns, these cries express more frequently desire than discomfort. The child then finds that the sounds bring a certain pleasure in the mere uttering, movements of the lips and tongue tickle and please him and sounds uttered amuse him. This is probably the parrot stage. The first sounds which we hear from the baby are the guttural and the labials.

One who has listened to many babies just beginning to talk remembers that "goo goo" and "boom boom" are among the earliest sounds uttered. Often parents take these sounds to mean a desire for water and the child then learns to associate water and the satisfaction which this gives to its needs with the production of the sound which it has made. In this way the baby makes sounds which are the real "baby talk" and at first have no meaning, being simply produced because they are pleasurable and easily produced. Later the child learns to associate the sounds which it has made with the things that are given it or shown it in response to its cries. The instinct of imitation in a child is active and those initial sounds are repeated which the child is able to articulate. Dentals and labials follow the guttural and vowel sounds. During this stage Ferrard says the child is like a parrot and a good deal of its learning at this time is like the process described by Thorndike and Kinnaman in the observed monkeys. There is no cognition or abstract thinking possible in the average child in its first year and a half or two of life, it learns simply by associating sounds with objects and persons seen, felt, tasted and smelled. When this step is carried further and the children are able to arouse auditory, visual, tactile and olfactory memories which already have been registered by this process, and to recall pleasurable proprioceptive sensations, then occurs a combination of these images, and abstract thoughts and concepts are expressed in speech. For the child to arrive at this point many stages of a gradually developmental sort are traversed and it is not until the power of abstract thinking and intelligence in its highest form is developed that we have speech in its broadest sense in the child. One need only recall instances in his own experience with children to see how great a variation there may be in the methods by which children are taught to associate and later to speak and think. In the production of human intelligence and speech there are at least five great afferent pathways which are used upon the basis of the instinctive necessity which the child has to express its desires, its needs and its emotional and instinctive states. These are the visual, auditory, tactile, proprioceptive and olfactory, and taste. Depending on the degree of attention which the child gets from its relatives and parents and the individual preference or accidental methods used by them, various pathways are used either almost alone or together in a fairly even proportion. The least variable, undoubtedly, is the olfactory.

The education of some children may be accomplished through the use of one or a combination of some of these pathways to the practical exclusion of the others. Some mothers and relatives will sit and talk and imitate the child's sounds and use the child's auditory mechanisms, others show the child things and use the apparatus for visual registration, while still other parents and relatives neglect their children and the babies have to learn by feeling, tasting, and by the proprioceptive senses, being enabled in this way to associate words and speech with objects and persons smelled, touched and tasted. Therefore one can readily see that the development of a regular pattern of speech "centers" in the brain is impossible in any considerable majority of individuals, because serious variations occur in the method of education and the synthetic production of intelligence and speech in the individual child. This undoubtedly accounts for the fact that it is impossible to follow in any great number of people the diagrammatic scheme of definite speech and intelligence "centers" for the combination in speech of auditory, visual, tactile and kinesthetic memories. Later, as the child begins to associate writing and reading with thought and speech, "centers" for the retention of memories of written words and words which have been read must also be postulated if this diagrammatic scheme is followed. If it were possible to subject all children to the same method of education and to put them through the same process during this period of development of speech and intelligence, it probably would then be found that certain definite areas of the brain which had to do with the functions of speech, had been developed after a more or less regular pattern. This will explain why the anatomical and pathological investigations which will be mentioned later have been so potent a factor in breaking down our purely diagrammatic conception of sharply circumscribed speech "centers." Since the variations occurring with the individual child in the use of the various special senses (including the proprioceptive as one of these) are so great for the production of simple associations, and later of conception and thought which find their expression in speech, the nature of the development in the brain of this function will also vary greatly.

The reason why anatomy and pathology have failed to clear up the problem of aphasia is that many have looked upon speech as a function entirely separated and distinct from intelligence, and have set apart certain portions of the brain for its exclusive control. If this were true, the anatomical association paths in the brain should show in every case definite fiber tracts and bundles leading through from the main visual or auditory registering areas of the brain to the other so-called "centers" in a perfectly definite pattern to certain definitely circumscribed groups of cells or "centers" the form of which would vary but little in different individuals. A consideration of the anatomy of these association pathways shows that this is not so. As Herrick [illegible] states: "All parts of the cerebral cortex are directly or indirectly connected with other parts by these association fibers so that no region can be regarded as the exclusive seat of any particular cortical function." Although much study has been devoted to these various association fibers of the cortex, a great deal of irregularity and inconstancy of pattern has been found. The only association fiber pathways which have any definite degree of constancy and which can only be made out in the gross by anatomical dissection after hardening of the brain in appropriate fluids, are the cingulum, the fasciculi-longitudinales inferiores and superiores, the arcuate fibers or as they are sometimes called, the fasciculi proprii and the fasciculus uncinatus. Various other association bundles are known but are inconstantly arranged and not always easily identified. Of course the patterns of the great association tracts through the corpus callosum have a fairly definite arrangement, as do those of the fornix and the commissures (smell). The short association fibers pass in great numbers from one convolution to the next and are either intracortical or subcortical. The long association fibers connect the various large association "centers" of the brain in the frontal, temporal, temporoparietal and occipital regions. The most marked of these bundles, the superior and inferior longitudinal fasciculi and the uncinate fasciculus, pass from and connect with each other the frontal and parietal opercula, the inferior parietal convolution, the occipital and the superior and middle temporal convolutions, the tip of the occipital lobe and the cuneus, and the forepart of the temporal lobe. Naturally if the processes of association in the development of intelligence and speech were continuously the same, a constant pattern findable anatomically would result and these association fibers could be followed directly to ganglion cells arranged in definite groups in definite parts of the brain. Actually no such arrangement exists in the vast majority of brains examined by anatomists, and in fact, it is impossible to follow these association fibers directly to any of the so-called "centers." In point of numbers the greatest system of these fibers are the intracortical and subcortical association fibers, known as the fibrae propriae and tangential fibers, which run in the cortical gray. It is impossible to follow these out and there is certainly nothing in the nature of a definite pattern in their arrangement and distribution. In fact, this would appear to be exactly what one would expect, for as Piersol says,[15] except for the immediate vicinity of the upper end of the Rolandic fissure, "the cerebral cortex at birth is unprovided with association fibers which have acquired their medullary coat and are therefore capable of functioning. Within the early months after birth, however, the myelination of these, as well as of other tracts, progresses rapidly, although this process is not even moderately completed until after the lapse of several years. Indeed there is sufficient evidence to believe that myelination of additional fibers continues so long as intellectual effort is progressive, the demands made by education and special mental exercise being met by a corresponding completion of additional association fibers." These facts so well stated and based on indisputable anatomical evidence would of course preclude the possibility of the existence of any sharply circumscribed, purely diagrammatic localization of speech functions in any group of "centers."

PART II.

Head,[16] P. Marie [17] and Moutier [18] have all called attention to the necessity of revising our conception of speech and associated problems. Recently James Collier [19] also voiced his sense of bewilderment as a result of the confusing manner in which this problem has been presented and expressed his dissatisfaction with the diagrammatic localization theory of aphasia and the "center" conception of the development of speech. He calls attention to the fact that these schemes of exact localization in the brain for the various components of speech and for the various types of aphasia are based more on theory than upon trustworthy pathological evidence. His objections and those of Marie and Head are based on a very exhaustive consideration of the pathological side of the subject of aphasia. One need only review the statistics of Moutier, Marie, Frankel and Onuf [20] to become convinced that our conceptions of aphasia are in a developmental if not a chaotic state. The diagrammatic speech "center" theories have no basis in the psychology, anatomy or pathology of the subject. These authors have presented their conclusions based on an investigation of a total of 408 cases of aphasia with autopsy. Marie and Moutier in 304 of these cases found that the meager clinical and pathological records preclude the possibility of drawing conclusions of value in at least 201 cases. In the remainder of their cases the lesions were localized sufficiently and the records and clinical histories were of such a nature that a fair study could be made of the pathological findings and certain fairly definite conclusions drawn.

Recent criticisms of our conception of this subject as outlined in these pages and elsewhere make it necessary that we draw attention to the invaluable work of Moutier published in his book on aphasia. This author examined all the autopsied cases of aphasia which were reported in the literature from the time of Broca in 1861 up to 1906. Certain extremely important facts are overlooked or not known by our critics. Broca's original contribution, which consisted simply of two observations and were a basis for the later diagrammatic conceptions of speech, were not only imperfectly studied cases clinically, but absolutely valueless pathologically. The brains in these cases up to the year 1908 had not yet been sectioned. Criticisms of this kind should not detract from the splendid work of a pioneer, but nothing must stand in the way of a proper presentation of our subject. Moutier found that of the 304 autopsied cases 201 were absolutely useless for any study. The clinical records in these cases were very meager. Many of the brains had been reported upon without the meninges having been stripped off and most of them were unsectioned, so that the real extent of any pathological lesions present could not possibly be detected. Of these 201 useless observations 26 gave not only insufficient data but in 175 the destructive lesions were so extensive that it would have been useless to draw any conclusions from these brains. There remained in Moutier's series of 304 observations 103 with fairly well localized areas of destruction. There were 19 cases which were apparently favorable to the conception that speech function is localized in the left third frontal lobe. Of these 8 were ~~cortical and~~ 11 ~~subcortical~~ lesions. On the other hand, in 57 cases aphasia was present and the foot of the third left frontal convolution was absolutely intact. In 27 brains the foot of the third left frontal was destroyed and no aphasia had existed in the patients at any time. In 4 of these cases there had been extensive trauma, 14 were cases of tumor, 5 of softening and in 2 there were lesions on both sides in the third frontal. In 2 other cases the injury on the left side had been produced surgically in right-handed individuals. Therefore in these 27 cases with injuries or disease of the third left frontal no aphasia was present. In the light of such careful pathological reviews as Moutier has made of this subject one must hesitate before committing himself to the theory of the exact cerebral localization of the speech function. Certainly medical men do not accept other teachings with the same readiness with which many have accepted this conception of aphasia.

Let us remind ourselves of the fact that we insist on 100% proof in the elaboration of Koch's law when it is applied to the discovery of a new organism. Yet here in a very important problem many men are able to satisfy themselves with a conception which is borne out by only 8 of 103 autopsied cases. To become impressed with the importance of Moutier's work one must read his book and see the minuteness of his method of investigation and report upon all this autopsied material. His consideration of this material is on the whole fair and certainly very exhaustive. The diagrammatic conception of aphasia rests upon observations made upon brains, many of which have not been sectioned and which have been subjected only to the most meager sort of anatomical investigation. Many of the brains came from patients who were undoubtedly advanced seniles or diffuse cerebral arteriosclerotics. Some of the observations were made in brains taken from patients who died from tubercular or other types of meningitis. There were ten cases of this type including cases of specific or tubercular infection and post-traumatic meningitis. In the tubercular cases Moutier (p. 85) comments amusingly on the peculiar phenomena discovered by some of the men who reported these cases, that in the presence of hundreds of tubercles nevertheless most of them according to these observers were found in the left hemisphere and the vast majority grew upon the third left frontal convolution. Why the greatest damage in the traumatic and infectious cases should be over this area is also wondered at. Of the 103 favorable cases in Moutier's study there were 50 cases of brain tumor either with operation or operation and autopsy.

In considering these cases Moutier agrees with Marie that to use the symptom of aphasia as a localizing diagnostic aid is impossible and a dangerous procedure. In most of these cases of brain tumor, many of which were reported by surgeons and not by neurologists, the author doubted the presence of real aphasia. He points out the fact that with brain tumors, especially frontal lobe tumors, various speech, intelligence, memory and thought difficulties are found, but extremely rarely is there any pure type of aphasia. He says after considering these autopsied tumor cases, that "there is not one case of tumor involving exclusively the foot of the third left frontal convolution in right-handed individuals which has ever caused aphasia. Many times this convolution has been completely destroyed by the tumor and no aphasia has resulted."

His discussion of this subject brings to my mind a recent personal case. This man, aged forty-eight, died at the Neurological Institute on the service of Dr. F. Tilney, and was operated upon by Dr. A. S. Taylor, who found at the operation a large mass which turned out to be a mixed cell sarcoma (report by Dr. L. Casamajor) involving the left temporo-sphenoidal lobe and running inward almost to the midline. The mass practically destroyed the first, second and third transverse gyri of the left temporal lobe, penetrated the Sylvian area and the Rolandic fissure in a massive way and pressed also on the foot of the left third frontal convolution. Yet when this man was examined on March 16th, 1920, only five weeks before he died, he did not present any of the usual types of aphasia. He spoke well spontaneously, read and understood what he read, saw and understood what he saw, had no paraphasia, alexia or agraphia and had a perfect conception of speech. The type of memory defect which he showed seemed to point more to an association difficulty which made it almost impossible for him to recall well. He said that he was born in London in 1871 on the 25th of December and that he came to the United States with a woman, but it was not until later that he said this was his mother. He said that he was six months old when he arrived here and stated that the mother died at the age of seventy-five, but when asked what she died from he said "I do not remember." The mother died four years ago but he did not know the exact date of her death. He was able to recall that the father died from an infection of the right leg about six years ago but was not able to give the date of the death or an understandable account of the cause of death.

With considerable effort he gave the names of his brothers and sisters, but in recounting their ages he was only able to say that he remembered there was a two years' difference between them but he could not tell their exact ages. He said L. is fifty-three, M. fifty-two, and then as to each of the other six he was unable to give the ages. Concerning his brother R. he said, "He is eighteen months older than I;" but even after considerable effort he was unable to say exactly how old R. was. He had a perfect memory for what had transpired recently and his retention of test phrases and numbers for short and long periods (from five to thirty-five minutes) was perfect. The main difficulties seemed to be the recall of even well-known names and events and their calculation in point of time. He seemed to have difficulty in arousing both visual and auditory memories, but particularly the latter. There was no difficulty with speech on the motor side. His only trouble was a left-sided headache, not at all severe, and a definite memory loss. He spoke and read well from dictation. There was not in any sense of the word a real aphasia when I saw him.

This memory defect corresponds with the type of finding that Mingazinni [21] has called attention to in tumors of the temporo-sphenoidal lobes.

I saw another patient at the Italian Field Hospital No. 227 at Lonigo, whose entire left frontal and the greater part of the temporal convolution of the left side had been shot away by a piece of shell. The cranio-cerebral defect was enormous, but unfortunately the records which accompanied the patient were so incomplete concerning this point that the exact area of brain involved cannot be definitely stated. The patient was a right-handed man and he came of right-handed stock, yet, excepting for an initial speech confusion of paraphasic type which lasted for about two months following the injury and the operation which followed, he had no speech defect whatsoever. At the field hospital where I saw him about nine months after the injury, I could discover no speech defect which exactly fitted any of the schematic types of aphasia artificially constructed by theorists. He had some sharply circumscribed memory defects. He had forgotten the names of a number of his boyhood friends, and was unable to recall the name of one of his sisters, but he wrote long letters home, was able to read aloud and was able to speak and write from dictation and soon reacquired the ability to name and recall his sister after much prompting; but he was never able to recall exactly certain friends and some incidents connected with their boyhood associations.

Of the 104 cases of Frankel and Onuf,[20] only 4 were found which seemed to confirm Broca's theory of the localization of motor speech; but they found at the same time 5 cases of motor aphasia in which the left third frontal convolution was absolutely intact and the subcortical association fibers going from and to this part of the cortex showed no trace of degeneration. If these facts were not sufficient to cast serious doubt on our theories of the exact localization of speech in "centers," one might find it necessary to call upon other evidence to destroy the diagrammatic conception of speech localization; for instance, the operations of Burckhardt, a German surgeon, might be recalled, who deliberately operated two insane patients suffering from noisy over-productive speech, attempting by his operations to produce aphasia and thus relieve this symptom. He removed 5 grams of the cortical gray from the foot of the first and second left temporal gyri and no word deafness resulted. Eight months later in the same patient he resected the top and foot of the left third frontal convolution and no agraphia or motor aphasia resulted. In another patient this surgeon removed a portion of the left supramarginal convolution and destroyed a considerable portion of Wernicke's sensory speech area and no aphasia resulted. In this last patient the left third frontal convolution was also resected. The patients continued just as voluble and full of noisy speech as before.

Added to this convincing evidence is the flood of literature which has appeared during the war, of severe injuries to most of the frontal cortex including the third left frontal, or the first, second and third left temporal, the supramarginal and angular gyri and also of the subcortex, of all of these so-called speech centers. In a number of cases no aphasia resulted, in a great many the aphasia or rather the speech defect was temporary, and speech was regained in a great majority of the cases.

Moutier discussed in detail the 19 cases which are apparently favorable to the theory of Broca concerning the dominance of the left third frontal convolution of the speech functions. As previously stated, only 8 of these 19 favorable cases were subcortical. He points out in great detail that of all the 103 cases analyzed by him only 4 show an absolutely circumscribed lesion limited to this area and in the other 15 cases favorable to Broca's theory other portions of the brain, particularly the Sylvian territory, were involved. In some of the 19 favorable cases no mention was made of whether the individual was right or left-handed. In these same cases the state of the meninges and of other portions of the gray matter of the brain was not mentioned in the autopsied reports by Christianson, McDonell, DePallet, Rosenstein, Kervey, Ord and Shaddock. In the cases reported by Chauffard and Rothery discussed in detail by Moutier there was a very considerable involvement of the island of Reil and the Sylvian area. Even after considering the 19 cases apparently favorable to Broca's theory, Moutier agrees with Marie that there is not a single case of aphasia in which the lesion found at autopsy has been sharply circumscribed and limited entirely to the region of the third left frontal.

The criticism has been made that our viewpoint in recent writings has not been backed up by pathological material, yet the fact that Moutier has examined every reported case up to 1906 and that his 304 cases added to 104 cases earlier examined by Frankel and Onuf make a total of 408 cases of studied autopsied material available for this discussion, seems to have been overlooked. Out of these 408 cases of aphasia with autopsy Moutier was willing to concede that 8, and Frankel and Onuf that 4 seem to confirm Broca's theory. Certainly the percentage of confirmatory material, which consists of a total of 12 out of more than 200 well studied cases is not a very high one.

Passing reference is made for the sake of completeness to the type of autopsy material which forms the basis for the diagrammatic conceptions of that problem which is so intimately associated with the aphasia problem, namely, the matter of apraxia.

Reference to the work of Liepmann,[22] Bonhoeffer, Marcuse and Pick [23] will immediately show the same fallacious pathological material which Moutier has criticized in his review. The original case reported by Liepmann had not only right-sided apraxia but motor aphasia with considerable cutaneous and deep sensory loss on the paralyzed side clinically, but pathologically it was found that all of the corpus callosum excepting the splenium contained scarcely a single nervous element. There were two foci in the white matter beneath the second and third left frontal convolutions where the callosal fibers enter the hemisphere, and there was a large subcortical cyst on the left side beginning under the lower third of the postcentral gyrus and running back through the parietal lobe to beneath the supramarginal gyrus, but not in the angular gyrus. Besides all this, there were foci of softening on the right side in the internal capsule and another in the supramarginal and angular gyrus and marked general arteriosclerosis. The cases reported by Bonhoeffer, Marcuse and Pick to illustrate types of aphasia and of apraxia were cases of senile dementia, and to attempt to localize the pathological findings was too much even for these upholders of a diagrammatic speech and apraxia center theory.

It is a well known clinical observation that aphasics improve with re-education. Frazier [24] has recently mentioned the fact that a number of aphasics caused by craniocerebral injury improved greatly by re-education. These aphasic patients necessarily go through the same methods of learning language that are used in childhood and connections are established between association "centers" in the cortex by pathways that must necessarily be different from those previously used and now destroyed by injury, tumor or what not. Therefore this observation alone would be enough to cast serious doubt on any notion that only certain parts of the cortex are capable of developing the power to control speech.

Incidents of recovery without aphasia or with only temporary paraphasic speech defects following these craniocerebral injuries or tumors involving areas of the cortex and subcortex presumably having to do with speech, have been recounted and published by Kennedy,[25] Neuhoff,[26] Cushing,[27] Frazier [24] and others. Recently Morse [28] in a discussion of autopsied brain tumors reported six occurring in the temporal lobes. In discussing these he states that "aphasic disturbances are frequently absent in tumors of the temporal lobes," and quotes Cushing as stating that with the exception "of the uncinate area the temporal lobe is a relatively silent region even on the left side." Of Morse's six cases with autopsy only one had what might be called sensory aphasia. These tumors involved all four of the zones into which the temporal lobe has been divided by Mingazinni in his study of tumors. These zones include the hindmost part of the second and third left temporal convolutions which were involved in at least four of Morse's cases. Mingazinni also mentions the fact that aphasic disturbances are frequently absent in tumors of the left temporal lobe involving the cortical area known as Wernicke's zone and in many cases involving also the subcortex of this area. I have personally observed one case of injury to the hindmost part of the temporal lobe cortex of the left side with sloughing of a considerable area of this cortex following an operation, which resulted only in mild paraphasic symptoms which entirely cleared up in two months. Many of Mingazinni's, Kennedy's and Morse's observations were in right-handed individuals, and my patient was also right-handed and came of right-handed stock.

Kennedy's work on the subject of stockbrainedness in the so-called crossed aphasias brings up the matter of speech and other skilled acts and their disturbance in aphasia and apraxia having their exclusive localization in the left cerebral hemisphere. It is impossible to escape the fact that whatever there is of truth in the conception of "centers" for speech has been dependent upon some evidence that injuries, tumors, vascular accidents and so on, involving the left cerebral hemisphere, are much more apt to give aphasic and apraxic prenomena than like conditions in the opposite side of the brain. While in great part the observations, first of Broca and later of many others, would give considerable stability to the conception of the left hemisphere as the leading one for speech and skilled acts, we believe nevertheless, that this dominance depends not on any special localization of these functions exclusively in the left side of the brain, but upon a reliable anatomical and phylogenetic explanation. The left hemisphere in the vast majority of individuals is the earliest used both by primitive man and the child. Primitive man protected his heart by carrying a shield in the left hand and developed an offense with the right. He has carried down to us the habit of listening and looking almost instinctively to the right side, and one need only walk through a dozen classrooms in which children are being taught to note how common it is to have them turn the right side of their face and the right ear towards the teacher. One will note also that all attempts at writing are instinctively made with the right hand in children and any variation from this rule is very early discouraged by parents and teachers. One need only note too that in the vast majority of instances, speakers will tend to move a little to the left of the platform so that listeners must turn slightly to the right in hearing. Speaking platforms and pulpits are more often placed a little to the left, possibly for the same reason. The baby in his crib, time and again, so frequently that it becomes unusual to have it otherwise, is so placed in the nursery that the parent or relative or visitor stands at his right and talks to him and plays with him in such a way that it is nearly always the right hand of the baby which is the earliest used. The baby must turn to the right to hear and see sounds, persons and objects. Children are handed things to grasp usually in the right hand and men instinctively salute and clasp hands with the right.

It will be recalled that the law of myelinization establishes the fact that this process throughout the nervous system is coincident with the establishment of function. At birth according to Piersol,[15] Edinger[29] and Strong[30] the myelinization of the cortex is limited almost entirely to the upper end of the Rolandic fissure, no other portion of the brain being capable therefore of function. This area includes the precentral and postcentral regions. Edinger and Strong further state that next in the order of myelinization are the association fibers of the brain, later portions of the cortex about the Rolandic fissures, then the visual areas and the auditory areas of the cortex. At birth therefore the only portion of the child's cortex which is capable of functioning is the motor-sensory area. Naturally if the child is stimulated to begin the learning of his skilled acts by the use of the right side of the body, particularly the oculogyric functions and those of the right upper extremity, then it follows that the left cortex will be stimulated to the earliest development. In a number of children, however, this early left-sided brain development is lacking and through force of circumstances, possibly accidental, the left side of the body, particularly the left upper extremity, is earliest trained and the usual development of the cortex is correspondingly influenced so that the right cortex may predominate over the left or develop apace with it. The matter of right or left-handedness cannot therefore be of too great significance, because in the child during the process of development of all skilled movements, thought and speech, all other things being equal, the amount of extra development and use which the left cortex receives cannot be so overwhelmingly great that the right cortical areas suffer too much by comparison. If the conception of association fibers and tracts in the cortex, anatomically and psychologically correlated, is true, then association tracts must occur in fairly equal proportion throughout all portions of the brain, right as well as left, without too much favoring of one side or the other. In exceptional instances, however, the factors of development in the child may be so influenced that the matter of left and right-handedness can play an important part.

Kennedy refers to the statistics of Stier [31] who found that in 5,000 soldiers only 4.6 per cent were left handed and that in 60 per cent of these a definite history of sinistral tendency in their forebears existed. This might very well be so, for if a parent's best hand were his left he would have a natural tendency to approach his child from the left and to play with the child's left upper extremity and show him things from the left and speak to him from the left, in this way developing the child's left oculogyric and cephalogyric functions and left upper extremity and thus his right hemisphere. The factor of imitation in children would also favor this. One can see from Stier's statistics however that this occurs only rarely, and in confirmation of this Liepmann's work is also important. Liepmann agrees that 95 per cent of all individuals are right-handed. In discussing this subject, this author denies that either one of the hemispheres is necessarily the dominant factor in the production of apraxia. According to him, the chief factors in disturbances of all skilled acts were to be found in disease, injuries or other destruction of the fibers going through the middle third of the corpus callosum, the immense commissural pathway for association of the sensory-motor portion of the hemispheres. Liepmann also called attention to the fact that out of 89 patients with left hemiplegia there were 5 who were aphasic, and out of 41 right hemiplegics, 14 were found who were aphasic. While aphasia undoubtedly occurs with lesions of the left hemisphere almost six times as frequently as this condition occurs in lesions of the right hemisphere, nevertheless all of us have seen aphasia develop so frequently in left hemiplegics in right-handed individuals that the theoretical factors of stock-brainedness in correlation with the conception that one hemisphere is necessarily the leading hemisphere for speech and skilled acts cannot be the invariably constant operative factor. Hughlings-Jackson [32] in his many papers frequently insisted that neither hemisphere has the exclusive function of the speech mechanism. The author together with many others has described clinically cases of aphasia with left hemiplegia in right-handed individuals who came of right-handed stock. I have seen five such cases in the past three years.

The whole matter of left-handedness and right-handedness, stockbrainedness with one hemisphere or the other leading, must have a developmental and anatomical basis in the way which is outlined above and cannot possibly be an invariable constant determined only by eugenic factors. There seems no doubt that in a great many individuals the left brain develops earlier than the right and therefore is concerned more intimately with all specially skilled acts, articulate speech being included; but variations both accidental and developmental occurring during the education of the child and in his adaptation to environment may alter this usual difference, reverse it entirely, or nullify it sufficiently so that the difference in development of one hemisphere over the other may be negligible. Only such a conception would explain the confusing findings in cases of aphasia and apraxia on the autopsy table and at operation. As three of the greatest neurological surgeons in this country have said to me in personal communications, time and again aphasia and apraxia as exact localizing aids for tumors of the brain have completely failed them, sometimes even leading to operations on the wrong side of the brain. On other occasions they have found at operation tumors and other destructive processes in parts of the cortex said to be speech and apraxia "centers" without the type of aphasia or apraxia to be expected existing in the patient.[33]

Without going further into this phase of the problem, it seems to me that we have succeeded in destroying to a considerable extent the validity of the theory of circumscribed centers governing the functions of speech, as these are said to exist in Broca's or Wernicke's or any other area. Having indulged in this destructive criticism and review one must now substitute a possible basis for the understanding of speech.

PART III.

This review of the subject and elaboration of the conception that speech and intelligence develop apace and are part of the same seat in the brain, is not merely a viewpoint, but it is an attempt to rationalize our conceptions of the subject. We do not wish of course to attempt to upset the already proved facts of association areas in the cortex for the retention of memories and concepts whether they are visual, auditory, olfactory, proprioceptive, kinetic, etc. It doubtless is true also that close to the area for the actual movement of the organs of speech there are more or less closely grouped association cells and fibers which are concerned with the movements of these in speech. But, that these purely motor "centers" are connected by many intricate and complicated association pathways searching out all parts of the brain, is undoubtedly also true. Only by the use of this elaborate and extremely complicated mechanism is the human being able to express his thought by speech, whether this be articulate, or by the assumption of attitudes or by a look or a change in intonation of the voice, or whether it be by writing, reading, singing, laughing or crying. It is probably true that in a small percentage of individuals the process of learning, with the development of intelligence and speech, has been so exactly similar, that in them certain lesions interrupting definite pathways may give similar defects in the process of thought expression in speech, and that in this very limited number the use of the words motor and sensory aphasia, agraphia, alexia, etc., as sharply limited aphasic defects depending upon the conception which those words imply, is justifiable on purely anatomical and pathological grounds. On the other hand, it is our belief that in a much greater number of patients the process of the acquirement of speech and intelligence has been so irregularly different and individualized that to talk of patterns and any longer to hold the diagrammatic conception of speech as a function localized in definite centers is impossible and unscientific and is in contradiction of all our anatomical and pathological data and our knowledge of the development of speech in children and articulate utterance in intelligent animals.

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CORTICAL WORD BLINDNESS

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The production of speech is the most complicated function of the human organism. The correlation of auditory speech memories, visual images of written symbols, and the patterns for correct laryngo-glosso-labial movements, requires an interrelation of functions far more intricate than the farthest reaches of our physiological imagination will allow us to follow. It is not strange, therefore, that the interpretation of the varied forms of aphasia have given rise to a great deal of theoretical speculation. Our knowledge of the subject is very meager, but that there are more or less definitely localized anatomic seats where the memory images of the different sensory impressions are perceived, seems to have been established. New lights can be thrown on the subject only by a careful clinical study of patients with aphasia and correlation with autopsy findings. Unfortunately, many of the cases are so complicated that complete studies cannot be made, nor the exact condition determined.

Word blindness is one of the less common forms of aphasia. Two types have been described. The first, called by Dejerine "pure word blindness," is characterized by an inability to read written or printed words but with the preservation of the ability to write. This form is usually accompanied by a right homonymous hemianopsia. The second, rarer type, combines visual aphasia with agraphia. Ordinarily there is no hemianopsia in this form as the lesion of the angular and supramarginal gyri is more superficial and does not extend into the optic radiations of Gratiolet. The case recorded below in its clinical manifestations corresponds to the second group and, although it is incomplete in that there has been no autopsy, it is believed to be of sufficient interest to record.

HISTORY

Patient, Mr. A. S., aged fifty-five; married; a bricklayer by occupation.

Chief Complaint.—Inability to read for past three weeks.

Previous History.—Has always been well except for occasional slight head colds. Syphilis is denied by name and symptom. There was one attack of gonorrhea twenty-five years ago which was cured in two months.

Personal History.—He takes two cups of tea and two of coffee daily and smokes one or two pipefuls of tobacco a day. Has never used alcoholic stimulants to excess, his average being about two or three drinks a year. Has never taken drugs or medicines of any kind. His sexual habits are normal. Sleeps well; appetite is good; bowels move regularly once daily; urination occurs three or four times a day with no nocturia. He has five children, all of whom are well. His wife had one miscarriage.

His education was received at a parochial school in England and consisted mainly of reading, writing and arithmetic. He has never had occasion to do much writing as it was of little use in his occupation. Occasionally he has been a time-keeper and has had to keep records of the hours the individual men worked. He has never had any difficulty in doing simple arithmetic and in writing enough to meet his needs. He was in the habit of reading the newspaper every day and has always done this heretofore without the slightest difficulty.

Present Illness.—About three weeks ago he came home from work feeling well. After supper in the evening he started to retire at the regular time, but on lying down, suddenly had a feeling as though he were smothering. He immediately sat up and the feeling was relieved. All during the night he was unable to lie down as the moment he did so the sensation of smothering and choking returned, while he felt fairly normal when sitting up. The condition was so severe that he did not attempt to lie down for the next three nights, but remained seated in a chair. On the morning following the attack he noticed that he could not read the newspapers. He could see perfectly, but could not read anything. He attended a moving picture performance, but was unable to read the titles although he understood the movements of the actors and the scenes they were portraying.

So far as he knows, he has no difficulty except in reading. He goes about town without any trouble, recognizing his friends and all that goes on around him. He can identify a good many letters of the alphabet, and by spelling each word aloud or to himself, he can read a little; but if the words are long he is unable to identify them. He believes that he can write but has not tried to do so since the onset of his illness. He is not in the habit of writing much, but has frequently written letters to his wife when absent from home. He has noted no trouble whatsoever with speech, never using the wrong word, hesitating or repeating. He can say the alphabet and can count. At no time has he noticed the slightest difficulty in hearing or understanding what has been said to him. He thinks quickly and clearly.

PHYSICAL EXAMINATION

The patient is a well developed and well nourished man of fifty-five who does not indicate that he has used alcohol to excess.

Head. Eyes.—Conjunctivae are clear; no petechiae or other abnormalities.

Nose.—No discharge or evidence of obstruction.

Ears.—Negative to external examination. No mastoid tenderness.

Mouth and Throat.—Teeth are in fairly good condition, though there is a moderate amount of repair. Pyorrhea is absent. The tonsils are small and not inflamed. The tongue is covered with a grayish fur.

Neck.—There are no enlarged lymph glands. The thyroid is not visible or palpable. The jugular pulse is visible and the auricular wave is absent.

Chest.—Well developed and symmetrical. The movements of respiration are normal and equal on the two sides.

Lungs.—There is normal resonance throughout both lungs. The breath sounds everywhere are of normal character. There are no râles.

Heart.—Normal in outline, the apex being in the fifth space, 9½ cm. from the midline. The first sound is rather muffled; the second sound is of normal character. No murmurs are made out. The rhythm is absolutely irregular. Pulse: Rate, 98; force and rhythm irregular.

Abdomen is rounded, slightly obese, soft, with no tenderness, masses or signs of fluid. The liver and spleen are not felt.

External genitalia appear normal.

Extremities.—There are slightly varicose veins in the calves of both legs, but no swelling of the feet or ankles.

Summary of Physical Examination.—The only abnormality found on physical examination is that of auricular fibrillation, there being, however, no signs of cardiac decompensation.

NEUROLOGICAL EXAMINATION

Abnormal Attitudes and Deformities.—Absent.

Gait.—No abnormality.

Coordination.—The patient is right-handed. Equilibratory: Standing with feet together, eyes open or closed, there is no swaying. Non-equilibratory: Finger-to-nose, pointing and pass-pointing, heel-to-knee-and-along-shin tests are performed normally. There is no adiadochokinesis nor dysmetria.

Skilled Acts.—There is no apraxia. Handwriting is difficult and will be discussed later. Speech: Conversation is normal.

Test Questions for Aphasia

1. *Can the patient speak spontaneously in ordinary conversation?*

There is no abnormality of spontaneous speech. His vocabulary seems to be better than that of the average in his occupation. He speaks clearly, with normal enunciation, never pausing for want of a word or using a wrong word.

2. *Can he enumerate and denominate objects?*

He is able to count fingers, coins, etc., correctly and has no difficulty in naming any pieces of furniture or other objects in the examining room or in the street outside the window. He says the alphabet, days of the week, months, seasons, and counts correctly and without hesitation.

3. *Can he speak from dictation?*

He repeats sentences which are read to him, without hesitation or mistakes.

4. *Can he read aloud?*

He was shown the alphabet and read the letters as follows:

M,	"M"	D,	"D"
Q,	"C or G"	J,	"J"
A,	"A"	E,	"E"
R,	"R"	I,	"I"
P,	"P"	F,	"F"
Z,	"Last letter of alphabet"	T,	"T"
O,	"C — G"	L,	"L"
W,	"T"	V,	"V"
B,	"B"	Y,	"Y"
S,	"S"	U,	"U"
N,	"N"	G,	"B; no, then G"
X,	"X"		
C,	"G"	H,	"H"

He is unable to read a single word without spelling it. He spells the letters aloud, thus rousing the auditory word center and Broca's center. In spelling long words, though the letters may be read correctly, he becomes confused and is either unable to recognize the word, or misreads it; for instance, *Manhattan* was read "Matron."

The following words were all spelled out loud and pronounced according to the sound of his spelling:

WE	"W E — WE"
BY	"B U — BU"
UP	"U B — UB"
IN	"I N — IN"
AT	"A T — AT"
DO	"F O — FO"
OF	"O F — OF"
HE	"H E — HE"
AS	"A S — AS"

BAD	"B A 4th letter of alphabet"
HIM	"H I L"
THE	"T H E — THE"
TOP	"T O Y — TOY"
GOD	"G O N — GONE"
SAID	"S A I D — SAID"
WORK	"G O U B — GOUB"
JUNE	"J U N E — JUNE"
WATER	"W A T E R — WAITER"
LAYER	"L A Y E R — LAYER"
QUILT	"G U I L T — GUILT"
JUDGE	"J U L G E — JULGE"
WAR	"T A V — TAV"
RAN	"E A N — EAN"
FOR	"F O R — FOR"
LAD	"L A B — LAB"
FARM	"F A B E — FIB"
VERY	"T E B X — TEBX"
LATE	"L A R E — LARE"
BRICK	"BRICE — BRICE"
YEARS	"U E A R S — WEARS"

In spelling out longer words he usually becomes confused and when he reaches the last letters, has forgotten the first ones, so that he cannot read the word. Sometimes he reads only the first two or three letters and then supplies the remainder of the word with anything that comes into his mind.

CLINIC	"C L I N I C—CLINIC"
EXCEPT	"E X C E P T —EXCELLENT"
TOWARD	"T O W E R — TOWER"
BEHIND	"B E H I N D—BLIND"
COUNTRY	"C O U N T R Y —CONSTANT"
INSTEAD	"I N S T E A D —INSTEAD"
UNDERWEAR	"UNDERWORLD"
TAXATION	"T A L A T I O N"
ANYTHING	"A L V G P N G"

GERTRUDE	"G R E A V E R"
FOREMOST	"FORREST"
METHODIST	(Unable to read at all)
NOTHING	"N O T H I N G —NOTED"

The following words and sentences were shown to him to see whether he would read the words spelled incorrectly in the same way that he read those spelled correctly. He did this in almost every instance, and in no case did he recognize any errors in spelling:

SOPE	"SOAP"
SOAP	"SOAP"
KRIST	"CHRIST"
CHRIST	"OUR SAVIOUR"
KRIM	"CRIME"
CRIME	"CRIME"
MANNHATAN	"MATRON, LADY OF THE HOUSE, SERVANT"
MANHATTAN	"THE SAME"

I HAV A BLAK KOW
"I HAVE A BLACK COW"
I HAVE A BLACK COW
"I HAVE A BLACK CALF"
INGLAND
"INLAND"
ENGLAND
"THAT'S THE SAME THING"
I LA BRIKS
"I LAY BRICK"
COM HER
"COME HERE"
GIVE ME A NIF
"GIVE ME A KNIFE"

When letters are traced on the palm of the patient's hand by the examiner, he names them as follows:

A	"A"	T	"U"
B	"B"	V	"V"
E	"D"	H	"W"
C	"O"	W	"T"
F	"P"	Z	"Y or U"
E	"Q"	Y	"X"
R	"U"	I	"I"
G	"S"		

Numbers that are traced on the palm of his hand in the same way are all correctly named; and when numbers are interspered with letters, he recognizes them all except the number 3 which he calls "a B without a front on it."

When the tip of his finger is passed over a surface forming different letters, he reads them in the following manner:

A	"A"	N	"N"
B	——	O	"O"
C	"N"	P	"P"
D	"E"	Q	"Q"
E	"E"	R	"R"
F	"Like E but not E, it is B"	S	"R"
G	"P"	T	"R or A"
H	"M"	U	"O"
I	"I"	V	"Last letter of alphabet
J	——	W	—
K	"H"	X	"X"
L	——	Y	"G"
M	"M"	Z	"R or U"

Reading of figures:

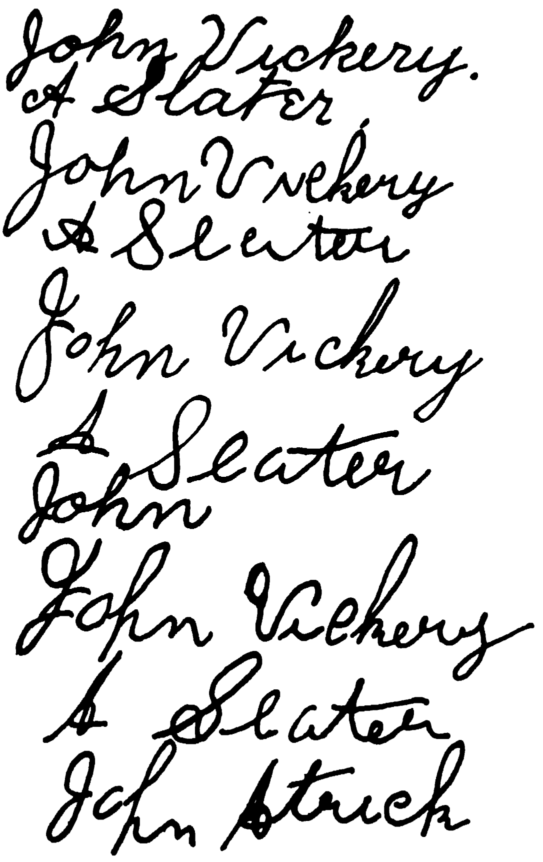
7	"Seven"
65	"Thirty-five"
489	"Four hundred and eighty-six"
3962	"Three thousand three hundred and sixty-four"
10602	"Two thousand six hundred and twenty."

5. *Can he read to himself and understand what he reads?*

He cannot read to himself as readily as he can aloud. All words must be spelled before they can be recognized. Any words that can be read are understood.

6. *Can he write spontaneously?*

His penmanship was never good and his occupation called for little writing. When away from home he wrote letters to his wife and children. Unfortunately, none of these letters can be obtained, and the only available writing that he had done before his present illness was in an old time-book. Figure 1 is a page from this book. Writing now is al-



John Vickery.
A Slater
John Vnekery
A Sleater
John Vickery
A Sleater
John
John Vickery
A Sleater
John Strick

FIG. 1. HANDWRITING PRIOR TO ONSET OF ILLNESS.

most impossible. When directed to write something spontaneously, he wrote the lines shown in Figure 2. An hour and a half was required to write these few words. The writing is almost illegible although a number of the words can be made out.

Upon being asked to spell certain words, he did so in the following way, the names at the end being the names of his own children:

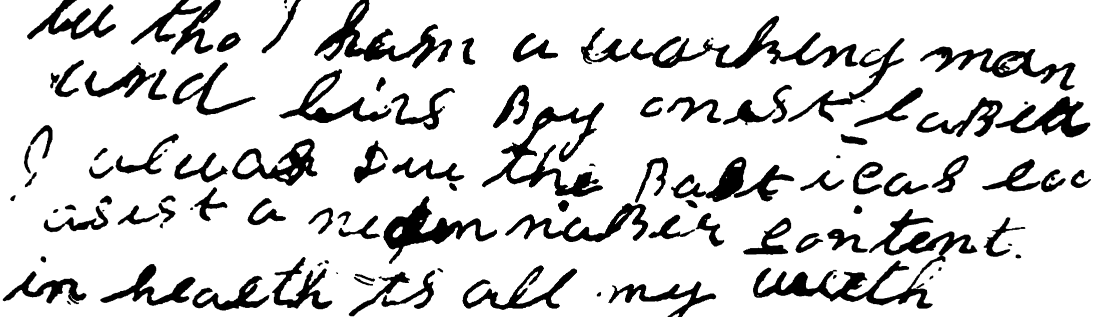
Bread	"B R E A D"
Tea	"T A"
Coffee	"C O F F I E"
Eye	"I E"
West	"W E S T"
See	"S E E"
Calf	"C A F E"
Few	"F E W"
Many	"M A N N E Y"
Hand	"H A N D"
Two hours	"T O W A U R S"
Gertrude	"G U R T R U D E"
George	"G O R G Y"
Edward	"E D W A R D"
Mabel	"M A B B L E"

8. *Can he write from dictation?*

He was asked to write the letters of the alphabet with the result shown in Figure 4. He was requested to make printed capital letters when possible, but if not possible any character that he could remember. After completing the alphabet he was directed to write "This is a fine day." His attempt to write this sentence is seen in the lower part of Figure 4.

9. *Can he enumerate and denominate in writing, e.g., write the numbers from 1 to 10, add a column of figures, and write the names of objects shown?*

He writes the figures 1 to 10 very quickly and easily. (Fig. 5.) The addition of two or three single figures of one denomination can



tee tho I ham a warking man
und hirs Boy cnask laBed
I wluas sue the Batt ieas lcc
asist a neomnaBer eontent.
in health its all my weth

FIG. 2. SPONTANEOUS WRITING.

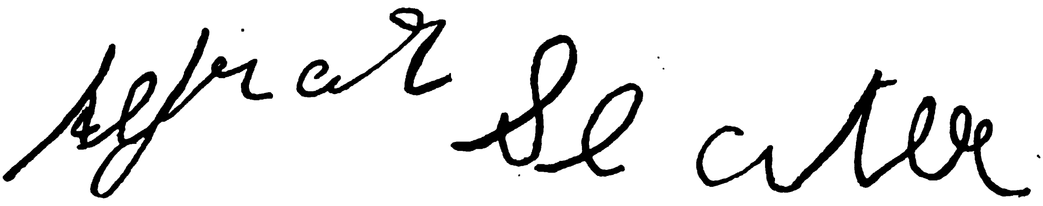


FIG. 3. WRITING OF HIS NAME WITH EYES SHUT.

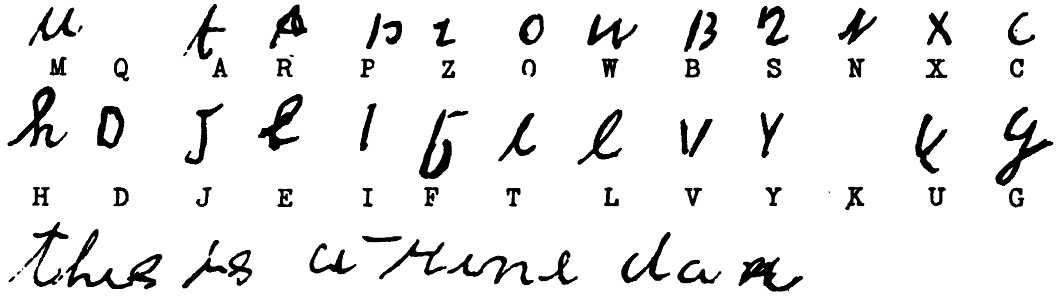

M Q A R P Z O W B S N X C
H D J E I F T L V Y K U G

FIG. 4. ATTEMPT TO WRITE ALPHABET FOLLOWED BY A SENTENCE.

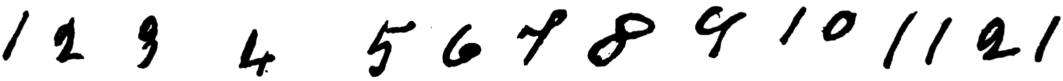


FIG. 5. WRITTEN FIGURES.

be done correctly, but more complicated addition cannot be done at all. He is almost unable to do any multiplication, failing in 2 x 10, 32 x 83, 28 x 83, though the last two examples were found correctly figured in the time-book which he had kept before his illness commenced.

In writing the names of objects, when shown a hat, pocket-knife and bottle, he laboriously wrote what appeared to be "hat, nife, bottel."

10. *Can he copy written and printed matter?*

He is unable to copy written matter. His copy of printed matter is shown in Figure 6.

11. *Can he hear and understand what he hears?*

He hears perfectly and understands perfectly words and sentences. Nor has he any difficulty in interpreting sounds, such as the tick of a watch, running water and footsteps.

12. *Can he see and understand what he sees?*

He sees perfectly and understands what he sees. On being shown pictures of animals, houses, furniture, flags, a barber pole, the three-balls sign of a pawn shop, a tennis racquet, etc., he recognizes all of them and understands their signficance or use.

13. *Is he aware of errors made in speech or writing?*

He makes exceedingly few mistakes in speaking, certainly no more than anyone of his education and station. He is not aware of errors made in writing and cannot read what he has written.

14. *Has he a concept of speech?*

He is unable to give the number of letters in simple words except by spelling them and counting the letters on his fingers. He does not understand what a syllable is.

15. *Does he misuse words? Is he ungrammatical out of proportion to his education?*

Words are not misused and he is not ungrammatical out of proportion to his education.

Reflexes:

DEEP	*Right*	*Left*
Jaw	1	1
Pectoral	1	1
Biceps	2	2
Triceps	2	2
Radial	2	2
Ulnar	1	1
Wrist	1	1
Patellar	2	2
Achilles	2	2
SUPERFICIAL		
Abdominal	Present	Present
Plantar flexion	Present	Present
Babinski and modifications	Absent	Absent

Muscle Strength.—There is no weakness in any group of muscles.

Muscle Status.—Volume, contour, consistency and irritability are normal. There is no myoidema.

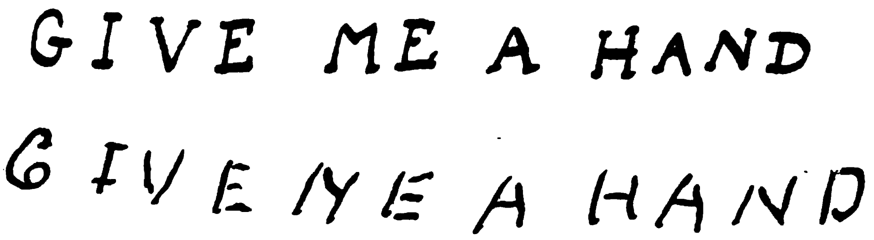

GIVE ME A HAND
GIVE /YE A HAND

FIG. 6. COPYING OF PRINTED MATTER.

Abnormal Associated Movements.—Not tested.

NERVE STATUS.—No observation.

GENERAL SENSORY.—There is no disturbance of touch, pain, temperature sensibility. Muscle tendon sense and stereognostic perception are normal.

CRANIAL NERVES AND HEAD. *Olfactory Nerve and Nose.*—No observation.

Optic and Oculomotor Apparatus.—Vision: right 20/40; left 20/40. Peripheral fields normal. Fundi normal. Pupils are equal, small, circular and react readily to light and on accommodation. There is no exophthalmos, enophthalmos, ptosis, strabismus, nystagmus or limitation of voluntary movement.

Acoustic Nerve and Ear.—There is no subjective disturbance and objective acuity is normal on each side.

Trigeminus Nerve and Mouth.—Movements of the jaw are normal. The corneal reflex is present on each side.

Facial Nerve and Face.—Taste not tested. There is no weakness in the movement of the upper or lower facial muscles on either side.

Glossopharyngeus and Vagus Nerves, Pharynx and Larynx.—The uvula is in the midline. The palatal and pharyngeal reflexes are normal. There is no difficulty with swallowing.

Spinal Accessory Nerve.—Normal.

Hypoglossus Nerve and Tongue.—Normal.

LABORATORY EXAMINATIONS

Blood Wassermann.—Negative.

Urinalysis:

Color—Light amber.
Specific gravity—1.015
Albumin—Very faint trace
Sugar—None
Microscopic—Occasional white blood cell. No casts or crystals.

SUMMARY

A right-handed man, age fifty-five, suddenly became unable to read written or printed matter. There was no paralysis, anæsthesia or hemianopsia. Intelligence was normal. There was no defect in speech, either spontaneous, repeated, or in singing. Understanding of spoken words was normal. There was no object blindness. Word blindness was complete, but most of the letters of the alphabet could be recognized. In reading letters mistakes were frequently made and the identification lacked assurance. He could not read words of even two letters without spelling them. Simple words whose spelling was phonetic were recognized after they had been laboriously spelled out loud. In longer words —those of six letters or over—he usually became confused and would supply the last part of the word with the first ending which came into his mind. For example, *except* was read "excellent"; *nothing,* "noted," etc. Spelling was phonetic, and written words spelled incorrectly but phonetically were pronounced according to sound. In some instances they were more easily read than when spelled correctly. Arabic figures were recognized much more readily than letters when occurring singly, but, were read incorrectly when combined to form a number of the third or fourth denomination. Chirokinesthetic images of letters were only exceptionally recognized, while those for figures were noted correctly and without difficulty. Spontaneous writing was almost impossible. The words were laboriously written out letter by letter. Many of the letters were illegible, some were left out, some repeated, and frequently the wrong letter was used. Writing from dictation was no better than spontaneous. Copying of written words was impossible, but copying of printed words was fairly successful. Writing of figures was but little impaired, and simple arithmetical addition and subtraction were possible, while multiplication and more complicated figuring could not be done.

This case belongs to the group of so-called cortical word blindness where word blindness is associated with agraphia. The lesion is probably located in the white matter of the angular and supra-marginal gyri, but not extending deeply enough to involve the optic radiation of Gratiolet.

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GENERAL PARALYSIS: PRESENTATION OF NECROPSY MATERIAL AND DISCUSSION OF THE NATURE OF THE DISEASE*

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It is not always possible to follow an interesting case of neurosyphilis from the time of apparent onset of symptoms, through periods of intensive antisyphilitic treatment, and check one's findings at the postmortem table. This, however, was done in the case to be presented before the conference. This case will also serve as an introduction to the discussion of general paralysis.

P. C., male, forty, divorced, white, came to the Vanderbilt Clinic on June 10, 1918, complaining of weakness, marked sense of tiredness, and of occasionally seeing double on looking to the left.

Family History.—Negative.

Past History.—He was born in this country. Had a normal infancy and childhood. Had two years of high school education. At sixteen, he went to work for a theatrical company, and had occupied himself since then in various fields of the theatrical business from a stage hand to a vaudeville actor. Married at twenty, but was divorced after living with his wife for only one year. He contracted syphilis in 1908. No skin lesions were ever noticed. He was treated for one year with powdered potassium iodide. In 1913 he was given inunctions and potassium iodide drops for one year, and at the end of this period his blood Wassermann reaction was negative. He continued with the inunctions and iodides, but in 1916 his blood Wassermann reaction was again positive. He was then given three intravenous injections of salvarsan and six intramuscular injections of mercury. He stated that in the latter part of 1917 his blood Wassermann was negative.

Present Illness.—Dates back vaguely to about one year before his coming to the clinic. He then began to experience difficulty in getting on with his act, and he also began to complain of weakness in his feet, "and of unsteadiness in his gait." He gave up his theatrical work and became a clerk in a store; could not keep at his work. Finally he was obliged to call upon his friends for financial assistance. A month ago he began to see double when looking to the left.

He was sent to the syphilology department for treatment. There, he presented marked ataxia; ptosis of the lids; absent K. J's; positive Romberg; visual acuity 20/20 in the right and 20/50 in the left; eyegrounds were apparently normal; pupils were unequal and irregular in outline, reacted sluggishly to accommodation and not at all to light. His blood Wassermann reaction was ++++. His spinal fluid was examined on several occasions and showed the following:

Date of Examination	*Cells*	*Globulin*	*Wassermann Reaction*	*Colloidal Gold*
8/17/18	7	+	++++ to 0.2 cm.	5555431000
6/11/19			++++ to 0.2 cm.	
6/25/19			++++ to 0.4 cm.	

There was no record of a mental examination. Because of the colloidal gold findings he was diagnosed as a case of paresis, and, in view of his physical signs he was finally regarded as a case of taboparesis.

He received the following treatment:

1. Twenty-one intravenous injections of of 0.4 gm. of arsphenamin from June 14, 1918, to August 24, 1919.
2. Eleven intraspinous injections of serum according to the Swift-Ellis method rein-

forced by 1/5 mg. of arsphenamin from August 30, 1918, to August 26, 1919.

3. Nine injections of mercury salicylate gr. 1½ from June 15, 1918, to August 30, 1918.

In the latter part of August, 1919, lumbar puncture resulted on three different occasions in dry taps. He failed to return to the clinic in September. During that month he began to lose interest in his personal appearance, seemed despondent and nervous, and expressed the idea that he was afflicted with softening of the brain.

On October 1, 1919, while at supper, he suddenly lost the use of his left hand. He was put to bed, and on the following day he was found in bed in a dazed condition, weak, and bleeding from hemorrhoids. He was taken to Bellevue Hospital, where he was described as being in general good health; pupils were unequal, the left greater than the right, sluggish reaction to light; tremors of tongue and fingers; increased knee jerks, and some weakness of the left arm and leg. Mentally he showed some elation and euphoria, inclined to be resistive, at times was restless and tried to get out of bed, though he was hardly able to stand. He showed memory defect, and had no insight into his condition.

He was transferred to the Manhattan State Hospital on October 8, 1919. There he showed ataxia, Argyll Robertson pupils, positive Romberg, absent knee jerks, definite speech defect and blood pressure of 130-80. His heart, lungs and other viscera were negative. Mentally he was at first depressed and later restless. He was mildly euphoric. Said that he could use his hands as well as ever. His stream of thought was relevant and spontaneous, was correctly oriented and gave fairly satisfactory data of personal identification. He did show slight memory defect, especially for the more recent events. He had no insight into his condition and his judgment was defective. His blood Wassermann was negative. His spinal fluid showed slightly positive globulin, no cells, and two plus Wassermann reaction to 0.5 c.c. and negative Wassermann to 0.1 c.c. of fluid. He was diagnosed as general paralysis, cerebral type.

He was transferred to an industrial ward on October 27th, to be developed as a worker. On November 12th he returned from exercise, had a chill, and temperature went up to 103° and he showed definite evidence of pulmonary congestion at both bases. He died on November 18, 1919.

NECROPSY SUMMARY

I. General Appearance.—Autopsy performed eighteen hours postmortem on the remains of a well developed, fairly well nourished man of about forty years. There was slight excoriation over the left heel and over the mid-dorsal region of the spine. There was considerable inequality and irregularity in the pupils.

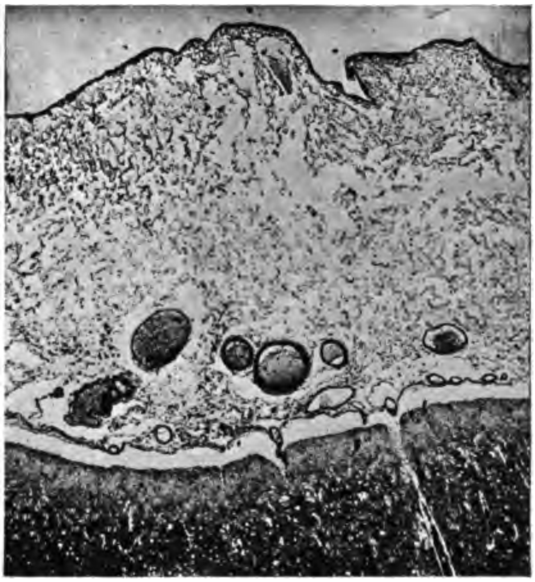

Fig. 1. (H. & E. X50) Frontal Pia. Note fibroblastic thickening, considerable edema, and occasional lymphoid and plasma cells.

II. Head.—1. *Calvarium* stripped with ease; bone appeared quite thin and atrophic; dura, when incised, allowed considerable spinal fluid to escape.

2. *Brain* weighed 1260 gm.; was of normal size and of somewhat increased consistency. There was only a moderate milky exudate over the anterior two thirds of the pia and most marked over the middle third. The posterior third of the pia appeared perfectly clear and glistening. There was moderate atrophy at the frontal poles and over the precentral areas, especially on the right side. The base of the brain showed very slight pial involvement, and mostly at the interpeduncular spaces, and at the ponto-cerebellar angles. The fourth ventricle showed an occasional granule, and the granulations were also seen in the lateral ventricles. The large vessels appeared smooth and were collapsed. Sections from the first frontal, gyrus rectus, precentral lobule and from the medulla were examined. The pia (Fig. 1) was decidedly edematous, showed a chronic fibroblastic thickening and contained a few lymphoid and a moderate number of plasma cells. The neuroglia layer was thickened and numerous neuroglia tufts ran into the subjacent cortex. There was a moderate loss of ganglion cells, especially in the gyrus rectus lobule. There was but moderate cortical disorganization, however, and while some sections showed quite a marked loss in ganglion cells, there were others that showed a fairly orderly and well preserved arrangement of nerve cells. There was a decided increase in the round glia cells. There was a distinct perivascular (Fig. 2) infiltration of lymphoid and plasma cells both in the cortical as well as in the medullary vessels. Occasional satellitosis was seen.

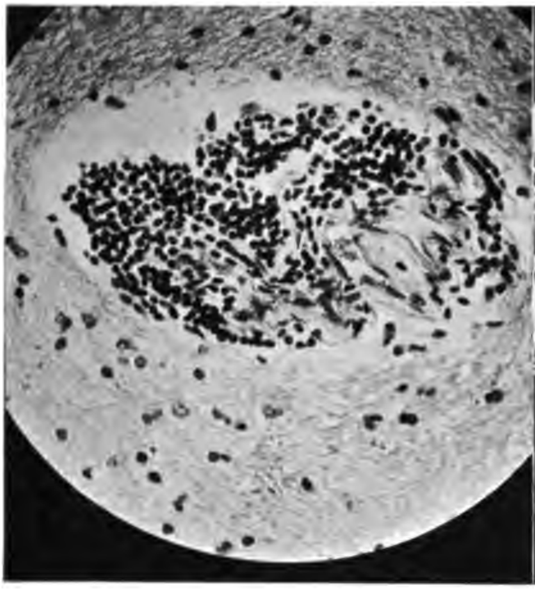

Fig. 2. (H. & E. X 400.) Deep Medullary Capillary. Note marked perivascular exudation of lymphoid and plasma cells.

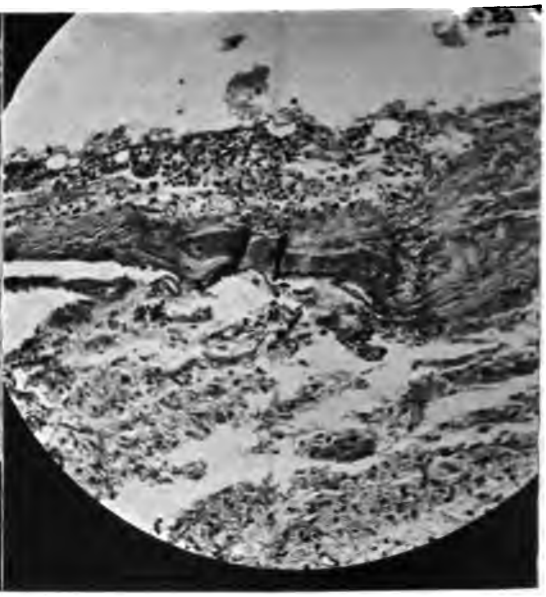

Fig. 3. (H. & E. X50.) Pituitary (nervous portion.) Note moderate lymphoid infiltration, and occasional plasma cell, in the capsule.

3. *The spinal cord* was removed. It showed dense adhesions between the dura, arachnoid and pia, and between the pia and the cord, so that it was difficult to separate the membranes from the cord in the lower lumbar region and at the cauda equina. In spots, the posterior columns appeared atrophic and somewhat retracted. Sections from the different levels of the cord showed a thickened pia and that was due to a fibroblastic reaction primarily, although there were in a few areas a few lymphoid and plasma cells. There was only a moderate loss of fibers in the posterior columns in the sections from the cervical and upper thoracic segments of the cord, but not sufficient to stamp it as a tabetic cord. There was a def-

inite marginal degeneration, best seen in the Marchi sections.

4. *The optic nerves* showed a few lymphoid and an occasional group of plasma cells between the fibers.

5. *The pituitary gland* showed nothing of significance macroscopically; microscopically it showed numerous lymphoid and plasma cells in its capsule (Fig. 3). Nothing remarkable was noticed in the cells of the gland itself.

6. *The internal ears* were opened and showed no pathological condition.

III. THORAX.—The sternum stripped with ease; there was very little retrosternal fat.

1. *Lungs.*—The right pleural cavity contained in its upper two-thirds recent and old adhesions which stripped readily on blunt dissection. Right lung weighed 635 gm.; was everywhere air containing; at its posterior two-thirds it contained considerable frothy fluid. The left pleural cavity was free from fluid and adhesions. Left lung weighed 485 gm.; showed chronic passive congestion.

2. *Heart.*—The pericardium contained a moderate layer of fat; the pericardial sac contained about 30 c.c. of straw-colored fluid. The heart weighed 335 gm. The muscle was of good color but somewhat soft and and flabby; thickness was normal; all values were compensating; the cusps were of a normal thickness and appearance. The coronary vessels were patent. Microscopic examination showed considerable congestion and increased fibrous tissue between the muscle fibers. There was a decided thickening in the capillaries.

3. *Aorta* was of normal caliber, generally elastic and pliable. Throughout the intima, however, there were a moderate number of pearly gray plaques, and thse were seen both in the thoracic and abdominal portion. Microscopically these were mostly fatty degenerations in the intima.

IV. ABDOMEN.—The abdomen was flat; the panniculus adiposis was of normal size and appearance; the peritoneal cavity contained neither fluid nor adhesions.

1. *Gastro-intestinal* tract showed nothing remarkable; the stomach was contracted; the large intestines were distended with gas and appeared thinned out; the small intestines showed nothing of significance; the appendix was of normal size and contained a few fecal concretions.

2. *Spleen* weighed 135 mg.; it was quite elastic and showed a definitely increased consistency. Microscopically it showed a definite fibrous tissue inflammation.

3. *Liver* weighed 1630 gm.; it was dense and elastic; it showed microscopically congestion and a definite increase in the fibrous tissue about the portal system. There was little injury to the liver cells.

4. *Pancreas* showed little of significance macroscopically.

5. *Kidneys.*—The right weighed 115 and the left 105 gm.; the capsules stripped with difficulty and left corrugated surfaces; the cortex was of normal size; the vessels were somewhat more prominent than usual. Microscopic examinations revealed a thickened capsule which sent numerous strands into the kidney substance. There was congestion of the capillaries and glomeruli. The ureters were patent. The bladder was contracted.

6. *The adrenals* showed thickened capsules and general congestion.

ANATOMICAL DIAGNOSIS

I. HEAD.—Diffuse meningo encephalitis (general paralysis). Marginal sclerosis of cord.

II. THORAX.—Right fibrinous pleurisy. Bilateral chronic pulmonary passive congestion. Aortitis.

III. ABDOMEN.—Diffuse splenitis. Diffuse hepatitis. Chronic nephritis.

SUMMARY

A man of forty years, of a negative family history and of a roving disposition, who had contracted syphilis in 1908, and had received treatment on and off for about ten years; who in 1917 began to show difficulty in getting along with his work, weakness in his feet, and unsteadiness in his gait; who come to the Vanderbilt Clinic in June, 1918, presenting marked ataxia, bilateral ptosis, absent knee jerks, positive Romberg, unequal and irregular pupils which did not react to light and only sluggishly to accommodation, whose blood serum gave a 4-plus Wassermann reaction and the spinal fluid showed 7 cells, 1-plus globulin, 4-plus Wassermann reaction and a paretic colloidal gold curve; who then received twenty-one intravenous injections of arsphenamin gm. 0.4, eleven intraspinous injections of salvarsanized serum, and nine injections of mercury salicylate gr. 1½ intramuscularly; who in September, 1919, began to show loss of interest in his personal appearance, despondency, and nervousness, and in October, 1919, suddenly lost the use of his left hand, becoming dazed and confused and had to be taken to Bellevue Hospital, from where he was committed to the Manhattan State Hospital; there he showed ataxia, Argyll Robertson pupils, positive Romberg, definite speech defect, restlessness, mild euphoria, memory defect, negative blood Wassermann, negative Wassermann to 0.1 c.c. of spinal fluid and only 2-plus to 0.5 c.c. of fluid, no cells and slightly positive globulin, and who at necropsy showed lesions of paresis, marginal sclerosis in spinal cord, right fibrinous pleurisy, chronic passive congestion of all viscera, and chronic nephritis, the immediate cause of death being right fibrinous pleurisy and pulmonary congestion.

DISCUSSION OF PARESIS

1. *Definition.*—As a result of the successful invasion of the brain by the spirocheta pallida, and consequent to the resultant parenchymatous degeneration and interstitial inflammation, there occurs a characteristic train of signs and symptoms, both physical and mental, causing a complete undermining of the somatic and psychic states of the individual, and producing a picture of a distinct disease entity known as paresis, general paralysis of the insane, dementia paralytica, or paretic neurosyphilis, depending upon the particular phenomenon of the disease which most strikes the student.

2. *Etiology.*—The demonstration of the treponema pallidum in the brains of cases of general paralysis has definitely [1] proven this condition to be a syphilitic process. Since this discovery, considerable work has been done in the attempt to solve many important and essential problems connected with the entire question of the genesis of this disease. It has been shown that many cases of primary syphilis may show involvement of the nervous [2] system. Most authorities agree that in the secondary stage of syphilis the nervous system is involved in from 18 to 70 per cent [3] of all cases. In fact, Wile and Stokes [2] venture to state that "in all probability every case of syphilis which reaches the secondary period has more or less involvement of the cerebrospinal axis." All who have had opportunities to study the spinal fluids [4] from cases of secondary stage syphilis have found alterations indicative of the participation of the cerebrospinal axis even in this relatively early period of the disease. As far back as 1906 Hoffman [5] was able to inoculate a monkey with syphilis by injecting into its eyebrow spinal fluid from a patient having papular syphilis. All evidence seems to point to an early spirochete septicemia, and to a localization of the spirocheta in this stage [6] and not in the late stages. And yet, in spite of the evidence of the involvement of the nervous system in a very large percentage of cases of syphilis which have reached the secondary stage, neurosyphilis represents only from 3 to 5 per cent [7] of all syphilis, and paresis, of course, would represent a still smaller percentage. There are several determining factors. First, there is evidence to suspect the presence of neurotropic spirocheta. Clinical proof is found in the fact that authentic cases have been reported where several men have developed paresis after they have lived with the same mistress at

different intervals. Experimental evidence is seen in the constant localization of certain strains of spirocheta in certain organs. Second, there must be a certain inherent factor which makes the nervous system of a given individual a fertile soil for the invading organisms. This is well stated by Mott[7] who says: "Pathological considerations as well as etiological facts, therefore, tend to show that another factor is required of a non-specific nature for the production of general paralysis, viz., conditions and habits of life tending to active metabolism of the nervous system involving mental excitement and stress, with a corresponding deficiency of conditions and habits tending to restoration of neuropotential, especially worry and insomnia; in fact all those conditions of civilization which produce neurasthenia, though not essential, are probably important contributory factors in determining the onset and progress of paralytic dementia." Third, there is undoubtedly a group of latent neurosyphilis cases which have never given any oddity in conduct or have never had occasion to be examined thoroughly by a physician, and in whom the disease has never been discovered, or it may have been detected accidentally.[8] Fourth, there is still an undetermined factor that must be at work in causing such specific anatomical alterations which produce the different types of neurosyphilis.

3. *Pathology.*—The anatomical alterations seen at necropsy and in the microscopic preparations in cases of paresis are quite distinct and definite in the vast majority of cases. The skull shows atrophy, and especially is this seen in the loss of marrow. Often one sees epidural hemorrhages. There is usually seen redundancy and puckering of the dura at the frontal pole, and this is due to the atrophy of the subjacent brain tissue. On incision of the dura and arachnoid, a large amount of cerebrospinal fluid escapes. The brain, in the vast majority of cases, shows well-defined signs macroscopically to enable the examiner to arrive at the correct diagnosis. The pia shows a milky exudate, most marked over the anterior two thirds of the brain, over the interpeduncular spaces, and at the ponto-cerebellar angles. The posterior third of the pia appears, as a rule, thin and glistening. The pial vessels are usually congested. In the ventricular walls and in the gyri recti there are seen pin-point elevations commonly spoken of as granulations or "sanding" of the ventricles. There is usually well defined atrophy of the anterior poles of the brain, and occasionally it is also seen in other parts of the cortex. The cranial nerves not infrequently show inequality and atrophy and thickened sheaths. The spinal cord in the vast majority of cases shows a thickened pia. Microscopically the lesions are distinct and clearcut in most of the cases. The pia is thickened, and shows lymphoid and plasma cells. The neuroglia layer is greatly increased, and tufts are seen running from this layer into the subjacent cortex. There is also a decided increase in the free glia cells. The cortex reveals disorganization —disorderly cell layer arrangements, loss of nerve cells, and all stages of cell degeneration. Occasional satellitosis and true neuronophagy are frequently met with. There is an increase in the capillaries, and budding branches are frequent. In the Vircho-Robin spaces are seen lymphoid, plasma and mast cells, the number, relative quantity and intensity depending upon the duration of the disease and the intensity of the inflammatory reaction. This perivascular cellular infiltration is seen both in the cortex as well as in the medulla. Rod cells are sometimes seen near the walls of the blood vessels and near the limiting glial membrane in the upper cortical layers. Sections through the walls of the ventricles passing through the granulations show glial hyperplasia with the resultant elevation of the ependymal layer; in places this layer may be split; rarely does one meet reduplication of the ependymal cells. Examination of microscopic sections from the cord often shows degeneration in the posterior columns, or around the periphery. The pia of the cord often shows thickening and lymphoid and plasma cell infiltration. Perivascular exudation of lym-

phoid and plasma cells are also seen in the vessels of the cord. Not infrequently one meets degenerative changes in the anterior and posterior roots. The pituitary gland quite often shows lymphoid and plasma cells in its fibrous capsule (this finding has been so constant in my material that it occurred to me that this must be responsible for a fair share of headache so frequently met with as a symptom in neurosyphilis). The optic nerves may show infiltrations of lymphoid and plasma cells between the nerve fibers. In properly stained sections one sees the spirocheta. They are found in the inflammatory cell infiltration in the perivascular sheaths, and near the nerve cells of the cortex. Mott[9] has recovered the organism from emulsions of the cortex in sixty-six out of one hundred consecutive cases of general paralysis. In from 10 to 15 per cent of cases, the posterior rather than the anterior half of the brain is most involved, and in these so-called Lissauer groups one meets clinically types that are characterized by focal signs of sensory, motor, and mixed nature, such as auditory and visual aphasia, apraxia, Jacksonian fits, etc. The other organs of the body usually show quite constant changes. The aorta most often shows pearly plaques throughout its entire course and linear scars in its thoracic portion; microscopically there are seen fatty degeneration in the intima and occasional loss of substance, infiltration of the media with lymphoid and plasma cells, and hyperplasia in the adventitia with perivascular cellular exudations in its vasa vasorum. The heart frequently shows fatty degeneration and brown atrophy. The lungs are usually the seat of a pneumonitic condition, most often of a bronchopneumonia which is responsible for the immediate death of most of the paretics. The large intestines usually have thin walls and the bowels appear greatly distended. Subacute or chronic nephritis, pyelitis, and pyelo-nephritis are often met with, while cystitis, in the hospital cases at least, is a very common finding.

4. *Symptomatology.*—While no two cases of paresis present the identical signs and symptoms, there is a certain uniformity in all cases of this disease not only in its somatic manifestations, but also in the resulting dementia. Physically the most striking alterations are seen in the pupillary responses, reflex disturbances, and incoordination, while mentally[10] the deterioration in the personality and the memory defect are the fundamental symptoms in the dementia. The pupils may be unequal, irregular in outline, and may vary in their reaction from mere sluggishness to a typical Argyll Robertson pupil or to fixed and rigid pupil. Gait may be ataxic. They may show positive Romberg and Fournier's tests. Their speech and handwriting are characterized by the omission of words, syllables and letters. Moderately rapid, medium and irregular tremors are present in the angles of the mouth, protruded tongue, and in the extended fingers. The deep reflexes are usually very active and often unequal on the two sides of the body. Occasionally the deep reflexes, especially the knee jerks, may be absent on one or on both sides. Often one finds pyramidal tracts involvement. Atrophy of some group of muscles is not infrequent. Sensory disturbances, while not at all rare, are not often found, and this is partly due to the poor cooperation on the part of the patient and to the tendency of the average paretic to minimize any symptom. Cranial nerve involvement other than the pupillary signs mentioned above are diminution in the visual acuity and occasional optic atrophy, extra-occular palsies, ptosis of the eyelids, deafness, difficulty in articulation of labials, change in the quality of the voice, difficulty in swallowing, and protrusion of the tongue to one or another side of the mouth. Convulsions are frequent in the later stages, and the patient is undoubtedly in a more advanced condition after each convulsion. Bladder and rectal disturbances are frequent in the advanced cases. The spinal fluid examination is an indispensable factor in arriving at the correct diagnosis as to the type of neurosyphilis with which one is dealing. The fluid usually escapes under considerable

pressure, and the true paretic hardly ever has a "puncture headache" and often his headache is relieved by a lumbar puncture. There is a pleocytosis, the number depending upon the intensity of the meningeal inflammation as well as upon the site of the inflammatory reaction; it is possible to have a normal cell count when only the meninges at the vertex of the brain are inflamed or when there is generally a mild inflammatory reaction in the meninges or when the process has petered out and there is present only the result of the acute process. Globulin is invariably present in practically every case. The Wassermann reaction is positive in practically every case of paresis. The colloidal gold reaction is of the "paretic curve" type, there being total discoloration in the first five or six tubes. In order to arrive at a correct diagnosis, it is imperative that all tests, globulin, pleocytosis, Wassermann reaction, and the colloidal gold test, be performed; and no spinal fluid may be considered as normal unless all these tests have been performed. As pointed out by Solomon,[11] there is a nonconcomitancy in the elements giving these reactions, and any one of them may be present or absent when the others are present. The correct diagnosis of the case presented was arrived at only after due evaluation of the different tests and especially in view of the colloidal gold reaction, in spite of the fact that the other tests were not in themselves pathognomonic of paretic neurosyphilis. The blood Wassermann is positive in from 95 to 97 per cent of the cases.

The dementia is characterized by the deterioration of the response of the individual to ethical, esthetic, intellectual and conventional standards which has hitherto stamped the individual as a social unit. There is a failure on the part of the individual to curb his aboriginal instincts and to subordinate his individual desires to those of social demands. There is a lack of evaluation of the various demands for adjustment and a consequent uniformity in the reaction, whether they be of the grandiose, depressed, or indifferent type. There is marked failure of memory. Retention is poor; judgment is gone, and insight into his own condition is hardly ever complete.

While the paretic is proverbially supposed to be quite grandiose in his mood, the depressed type is not infrequent. The depression is entirely different from the depression of a manic depressive individual. While in the latter there is still present a remnant of possibility and even probability in the imaginary causes of his depression, and we can even feel and understand his reaction; in the paretic, on the other hand, there is hardly a vestige of possibility in the imaginary cause, it is so improbable, silly and inconsistent as to make one immediately grasp the demented and deteriorated background into which are woven the patient's delusions. The agitated type is usually the most acute type and the one having the fewest remissions and running the most rapid course.

While the clear cases are easily detected even by the novice, the more unusual type of case offers considerable difficulty even to the specialist. Many "unusually interesting" cases of psychoneurosis, manic depressive psychosis, cerebral arteriosclerosis, etc., have turned out to be cases of paresis after the performance of thecal puncture. Vividly do I recall a series of chronic manic depressives who have been studied quite intensively by an eminent psychiatrist, and who turned out to be cases of general paralysis because of the positive spinal fluid findings in 90 per cent of these cases. The time is fast approaching when every hospital case will have a serum Wassermann performed, and every case with unexplainable pupillary disturbances or positive blood Wassermann will be subject to a spinal puncture.

The course of the disease is quite chronic, but the average paretic hardly lives more than five years after the onset of his distressing symptoms. The disease is characterized by remissions, and many a paretic is able to return [12] to his former sphere of activity. It is almost certain that no case in the stage of remission is as well as before the onset of the disease, and they usually fail to show that acuteness of reason and judgment which is necessary for the proper management of one's affairs. These remissions are biologic phenomena and are explained by Ehrlich [18] as manifestations of antibody formation. That they are not due to any change in the pathological picture is shown by the fact that cases coming to autopsy, having died of some intercurrent infection during a remission, show the usual findings of general paralysis. They are therefore not at all dissimilar to cases of pneumonia at the time of the crisis when there is no change in the anatomical picture but there is tremendous alteration in the clinical manifestations.

5. *Treatment:* Prophylactic.—It has been shown that there is an active septicemia in the early stages of the disease and the brain is invaded in the late primary and early secondary stages. If a real sterilization is expected, this is the time when active treatment should be done, as it is at this time that the spirocheta are most accessible to the action of medicinal agents. Intensive intravenous salvarsan administration, combined with intramuscular mercury injections, and if spinal fluid findings combine with clinical signs in pointing to an active and early involvement of the cerebrospinal system,[14] intraspinous treatment should be resorted to. No case of syphilis should ever get away from the proper supervision of the family physician. A yearly blood Wassermann and spinal fluid examination on most if not on all cases of syphilis would tend to minimize the frequency of the disease and would surely lower the 15 per cent of admissions to the State Hospitals for which neurosyphilis is responsible.

Specific Treatment.—In order to understand the rationale of the therapy it is essential to consider a few well established facts. It is well known that in disease the choroid plexus may become permeable to certain drugs and antibodies to which it was impermeable in health. It has also been shown that [15] irritation of the subarachnoid space with any sterile foreign body will greatly enhance the permeability of the choroid plexus. Mere spinal drainage [16] was found not to be sufficient to increase the permeability. It must also be remembered that those neurones which have undergone degeneration [17] have been lost to that individual, but the inflammatory reactions in the interstitial tissues and meninges are subject to regeneration and therefore might be spared. Furthermore, if the process could be checked from spreading, much would be accomplished. Therefore, in view of these facts, we believe that every case of general paralysis should receive the benefit of intensive intravenous salvarsan therapy combined with intramuscular injections of mercury and, in most of the cases, intensive intraspinous treatment. One who has had occasion to study these cases in the State Hospitals realizes the complete physical and mental destruction caused by this disease, and is sympathetic towards anything that would hold out a ray of hope for the amelioration of this condition. And results are constantly being reported. Intensive intravenous treatment has yielded a number of successful results in the hands of Southard and Solomon.[18] The work of Fordyce [19] is known to all and needs no comment. The rationale of treating paretics is well pointed out by Strauss and Kaliski,[20] though they differ in the method of treatment. These authorities differ on the frequency of the need for intraspinous therapy, although a closer study of their reports leads one to believe that there is more agreement upon the type of paresis that should receive such therapy than one is apt to think upon superficial perusal of their papers. One thing, however, is certainly well established, and that is this: Any mode of treatment that goes further than the intraspinous therapy is not the mode of choice even in the most advanced types of cases; and such methods as the subarachnoid and intraventricular injections of salvarsan are to be discouraged. We have had occasion to examine postmortem the organs of a case which had been treated by the intraventricular injection of arsphenamin and we found [21] that there was not only no favorable impression made upon the pathological process but there was greater cortical disorganization in the hemisphere which had received the salvarsan than in the other one.

Other methods of treatment, such as re-educational, hygienic and other corrective measures that have been found valuable in other diseases are quite as applicable to paretics as to other sick people and should be judiciously employed in treating this disease. Complicating conditions, such as cystitis, nephritis, etc., should receive the proper mode of therapy whenever they are detected.

In conclusion, we are indebted to Prof. F. C. Wood for the photomicrographs and to Dr. M. B. Heyman for permission to present the case before the conference.

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BULBAR AMYOTROPHIC LATERAL SCLEROSIS

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The two cases about to be reported present a few unusual features which are not commonly encountered in the course of amyotrophic lateral sclerosis. Both showed bulbar symptoms early in the course of the disease, Case II in fact beginning with them. Case I showed a myotonic symptom which is rarely or never seen in this form of amyotrophy, a fact which originally led one examiner of the patient to make the diagnosis of myotonia atrophica. Case II was further unusual in that it showed extremely rapid progress. The cases also illustrate a few well-known facts about pyramidal tract involvement in this disease. But before entering into a discussion of the salient features it may be well first to detail the clinical histories and physical findings.

CASE I. C. P., age forty-five, male, married, shoemaker by occupation, Jewish, born in Poland, was first seen at the Vanderbilt Clinic[1] (History No. 26981) December 29, 1919. At that time he complained of pain in the back of the neck, pain in the abdomen on bending over, inability to use the hands as well as formerly and difficulty in speaking. The condition began gradually in May, 1919, with weakness of the right index finger, which had rapidly grown worse. At first he noticed clumsiness of the right index and other fingers of the same hand when folding linings of shoe tops. Within three months the condition had grown so bad that he was compelled to give up his work. Soon the left hand became weak and he found difficulty in buttoning his clothes or doing ordinary skilled acts with the fingers of both hands. Very early in the course he found difficulty in voluntarily opening the right hand on flexing the fingers or making a fist, and he had to straighten the fingers out with the other hand. Little by little he began to experience dull, aching pains in the back of the neck, particularly on holding the head in an upright position for any length of time. Fatigue increased the pain and reclining relieved it. The patient also complained of similar pains in the abdomen, especially at the waist line, on stooping down or elevating the legs. Recently he found difficulty in speaking, claiming that he gets tired and that after a few minutes he has a feeling of pressure and weakness in the throat.

The past history is not very relevant. His habits are fair, he smokes fifteen to twenty cigarettes daily, only occasionally takes alcoholic drinks, he sleeps well, his appetite is poor, his bowels constipated, his urinary apparatus is normal. He never had any injuries, never was operated on, denies venereal diseases, thinks he had an hemoptysis at the age of thirteen, but no subsequent pulmonary symptoms. His wife miscarried once, ostensibly after an injury, and never conceived again. Since the onset of the illness the patient has lost twenty-five pounds.

On physical examination the patient betrays no abnormal attitudes or deformities, except in the hands (the right more than the left), which begin to look like "*mains en griffes.*" The gait is normal, there is no true disturbance of coordination in the upper or lower extremities although the finger-to-nose test is clumsily performed owing to the weakness of the fingers. Skilled acts are performed with great difficulty or not at all. Writing is preserved, but the lines are ragged. There is an irregular tremor of the fingers, medium in amplitude. There are myokymic twitchings in the muscles of the upper extremities, and possibly of the face and tongue.

The Deep Reflexes.—Jaw jerks lively and equal, about + +, the biceps, triceps, radial, suprapatellar and patellar very lively, about + + +, and equal, the ankle about + +. The ciliospinals are present, the abdominals absent, the cremasteric present R > L. No Babinski or any of its confirmatory signs.

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[1] Both this patient and the next were subsequently admitted to the service of Dr. Sachs at the Mt. Sinai Hospital for further study.

The patient rises with difficulty from the recumbent position. There is weakness of the arm muscles in abduction, weakness of flexors of the forearms and wrists, marked weakness of extensors, flexors, abductors and adductors of fingers; impairment of dorsiflexors of foot. The weakness on the right side is generally more marked than on the left. There is atrophy of the interossei of both hands, the thenar and hypothenar eminences; there is flattening of the forearm muscles and the pectorals. The muscles feel soft and are hypotonic and are markedly hyperirritable. No myoidema. The electrical reactions show hypo-excitability to both the galvanic and faradic currents—slow contraction and relaxation—but no reaction of degeneration. Marked myotonic symptom in the right hand.

There are no abnormal associated movements.

Sensation is intact throughout the body.

The Cranials.—Vision, fields and fundi are normal. The pupils are equal, regular, respond promptly to light and convergence, there are no ocular palsies, no nystagmus, possibly a slight divergent strabismus. Hearing is intact. The motor fifth is normal (?), there is flattening of the masseters, the corneal reflexes are present, the teeth and gums are miserably neglected. No facial palsies. Uvula normal, palatal reflex present, the tongue movements are normal; it possibly shows fibrillations, but there is some bilateral atrophy.

General Systemic Examination.—Negative. Blood Wassermann is negative.

The diagnosis at this time was myotonia atrophica because of the very marked myotonia of the right hand and the atrophy of the muscles. While there is some justification for considering this diagnostic possibility, it is obviously wrong. Myotonia atrophica really belongs to the group of muscular dystrophies wherein the myotonic symptom precedes or accompanies the muscular atrophy; it occurs in young people, frequently is familial, the atrophy is symmetrical and there are no fibrillary twitchings. The disease is in the muscles; the lesion is not in the central nervous system.

The patient was not examined again for six or seven months. When the writer saw him in July, 1920, he complained of marked weakness of the upper and lower extremities, wasting of the limbs, difficulty in speaking, swallowing and clearing of the throat. He had a distressing, hacking (laryngeal) cough. He had lost considerable weight—fifty or sixty pounds. There were no urinary, visual or auditory disturbances. Apparently the disease had progressed very rapidly—fourteen months since the onset.

To avoid repetition only the findings of diagnostic importance will be given.

There is marked fibrillation in the muscles of the neck, shoulders, extremities, trunk, tongue and possibly face. There is atrophy of the interossei, thenars, hypothenars, forearms, pectorals, shoulder muscles, face, masseters and tongue. Marked loss of power of extension of distals and proximal phalanges, and of the wrist. Weakness of flexors, abductors and adductors, of the fingers, flexors and pronators of the forearm, slight weakness of all the muscles of the shoulder girdle, weakness of the neck muscles. The right side is generally more involved than the left. The atrophy and muscular weakness in the muscles does not follow peripheral nerve distribution. Marked myotonia is present in the right hand in that the patient can flex the fingers to make a fist but cannot open the hand voluntarily or without very great difficulty. After careful observation the conclusion was reached that the condition was a true myotonia and not due to greater weakness of the extensors over the flexors of the fingers. Electrically the atrophic muscles showed absent faradism, marked hypogalvanism but not yet polar inversion, that is the qualitative changes of a beginning R. D.

The deep reflexes were universally increased: bilateral jaw clonus, all the upper and lower tendon reflexes were about 3 to 4 +. The upper abdominals were barely present, the lower absent, the right cremasteric absent, left diminished. Still no Babinski. Of the cranial nerves the motor fifth showed atrophy, the facial possibly; the tongue was markedly atrophic and could not be protruded well. The palate moved only moderately well. The larynx was not examined. Very extensive pyorrhea. The heart and lungs were normal. Blood pressure 139 S, 100 D. Urine negative. Blood and spinal fluid Wassermann negative, no cells or globulin in the spinal fluid. Blood count normal.

The patient received vaccine made from the flora in his mouth, but he became progressively worse. The condition has progressed so rapidly since, that a fatal termination is at the present writing apt to occur almost any day.

There seems to be no doubt that we are dealing with a case of amyotrophic lateral sclerosis. The condition began with nuclear atrophy of the small muscles of the hand, rapidly progressed to bulbar involvement and early compromised the pyramidal tract, as evidenced by hyper-reflexia, in some parts to the point of clonus. This hyper-reflexia is particularly significant because of its occurrence in the wrist, forearm and jaw where the atrophy was most marked. The absent superficial reflexes, too, point to pyramidal tract involvement. The salient feature, however, is the myotonia, and so prominent did this symptom appear that the original examiner was led to the erroneous diagnosis of myotonia atrophica.

Case II. S. W., age forty-six, female, married, Jewish, born in Russia, doing housework; was first seen by me in my office on May 29, 1920. Her chief complaints were difficulty in speaking, tremor of the fingers, stiffness of the tongue, inability to catch the breath on sipping liquids, nausea, inability to belch, choking feeling in the throat, difficulty in chewing, numb feeling in the left ear and a frozen sensation in the head.

The disease began gradually in February, 1920, with difficulty in speaking and has grown worse to date. On one occasion liquid regurgitated through her nose. She had no pains, no vomiting, no real difficulty in swallowing. Occasionally she feels better on resting the voice for an hour or so after speaking, although she does not seem to be better in the morning. She seems to have had chills at the onset of the illness, does not think she had fever, but insists that the disease had its origin in a cold. She had no visual, auditory or urinary disturbances. She never received an injury, was not previously seriously ill. Habits are good, sleeps well, appetite is poor, bowels constipated. Menses ceased in 1918, were previously regular. She complains of some dyspnea.

She had eight living children, five of whom died of various children's diseases, and one miscarriage. The family history is of no importance.

Physical Examination.—The voluntary motor system showed no abnormal attitudes or deformities, no disturbances in gait or coordination, skilled acts were well performed. The speech was distinctly bulbar in type, had a marked nasal twang and was particularly poor on labials and linguals. There was a coarse, regular, moderate tremor of the fingers while at rest and a similar tremor about the mouth. A few fibrillations were visible in the upper lip and tongue.

Deep Reflexes:

	Left	*Right*
Jaw	4	5 (clonus)
Pectoral	4	4
Biceps	3	4
Triceps	3	4
Radial	3	3+
Ulnar	2	3
Wrist	2	3
Suprapatellar	3	4 (clonus)
Patellar	3	3+
Periosteal	1	1
Achilles	3	4
Ankle Clonus	0	p

Superficial Reflexes:

	Left	*Right*
Ciliospinal	?	?
Supra-umbilical	1	1
Supra-pubic	0	0
Upper lateral abdominal	0	1
Lower lateral abdominal	1	1
Plantar flexion	1	1
Babinski	0	0
Oppenheim	0	0
Chaddock	0	0
Gordon	0	0

Muscle Strength.—Some generalized diminution of strength, almost hemiplegic in type, on the right side, rotation of the head to the left is impaired. Marked weakness or almost impossibility of moving the jaw to the right or very much from side to side. No atrophies noticed. Electrical reactions in the facial muscles were normal.

All forms of sensation were normal throughout.

Cranial Nerves.—Smell, normal. Vision showed O. S. 20/200, O. D. 10/200. The fundi showed slight blurring of the disks, the margins were not clear, veins well filled, but not tortuous (the refractive error possibly accounting for the picture). The pupils were dilated, unequal, the right being 6 mm., left 5 mm., they were regular, reacted promptly to light and convergence. No ocular palsies, no mystagmus; the palpebral fissures were unequal R > L; von Graefe present. Hearing was normal. The motor fifth showed weakness of the pterygoids, that is the lateral movements of the jaw. Both corneals diminished, R > L. Gums almost foul with pyorrhea. There was questionable weakness of the left side of the face. The uvula was in midline, the palate moved poorly during phonation, the palatal and pharyngeal reflexes were present, the voice was nasal. The tongue could not be protruded well nor moved laterally to each cheek, the movement to the left being poorer. When it was a little protruded it was in the midline. Trophic condition: no atrophy, slight fibrillations on the sides; very coated.

The patient is very emotional and cries readily, possibly significant of bulbar involvement. General systemic examination was negative. There was marked dermographia. Blood pressure 110 S. 80 D. Urine negative. Blood and spinal fluid Wassermann negative. Spinal fluid showed no globulin and few cells. Blood count normal.

About two weeks later the fibrillations in the tongue and about the lips became very marked, and they were readily discerned also in the hands and forearms. The deep reflexes became, if possible, even more lively and the superficial reflexes began to diminish and to be elicited with difficulty. Subsequent examination showed very rapid downward progress. The bulbar involvement is more marked. The face and tongue show atrophies and the fibrillations have greatly increased. The patient is at present in very poor condition, frequently is confined to bed and to the house for days at a time.

The diagnosis of bulbar amyotrophic lateral sclerosis seems to be justified in this case on the evident signs of involvement of the bulbar nuclei and the pyramidal tracts. The unusual features are the primary involvement of the bulb, with downward progress into the cord and the very rapid course of the disease.

Both cases made very rapid progress, though the first followed the more usual course. Such quick progress is quite characteristic of cases which exhibit bulbar symptoms, those in which the affection is more limited to the cord lingering on for years and years before terminating fatally. The writer once saw a patient who showed marked spasticity of both lower extremities with increased deep reflexes of the upper, who for a long time was looked upon as a rare example of pure lateral sclerosis. It was only later in the course of the disease when he began to show fibrillary tremors in the muscles of the upper extremities and still later atrophies that the true condition was recognized.

The spinal or protracted forms of amyotrophic lateral sclerosis in reality belong to a transitional form of the spinal or nuclear amyotrophies together with the so-called Aran-Duchenne types or even the chronic forms of poliomyelitis. Indeed many cases, contrary to the last one cited, begin with atrophies of the hands, etc., and only later develop pyramidal tract signs. The rapidly progressive form of amyotrophic lateral sclerosis always shows bulbar involvement. Such cases constitute a special clinical group, though pathologically they are part of a single picture and should not be separately classified.

The absence of the Babinski sign in all the three cases mentioned seems to be characteristic of amyotrophic lateral sclerosis, despite the evident involvement of the pyramidal tract. Recently Bing called attention to this in a study of the Babinski phenomenon and its various modifications and mentioned eight or nine cases of amyotrophic lateral sclerosis, some of which came to autopsy, in which the sign was wanting. Why this should be so one cannot say, but the fact remains that the absence of a Babinski should not militate against the diagnosis of pyramidal tract involvement in cases which show other signs of its involvement (spasticity, lively reflexes, absent abdominals) particularly when nuclear atrophies coexist.

In view of the unbridled, faddist tendency in recent years to attribute every disease of the body to focal infection I call attention with a good deal of trepidation to the fact that both patients suffered from very severe pyorrhea alveolaris. Whether the pyorrhea was the result of the inability to chew and speak properly and clean the mouth because of bulbar involvement or whether the former had a possible causal relation to the latter I do not know. The fact is merely mentioned because the involvement of the gums was extreme. In Case I a vaccine was made and tried on the patient with no success, the second patient did not submit to the injections of vaccine made from her own pus.

The myotonic sign in the first patient is very unusual and difficult to explain. One is not accustomed to encounter such a symptom in amyotrophic lateral sclerosis. That it was a genuine myotonic sign is not open to doubt. Several observers confirmed it. The muscular involvement cannot account for it. Why a sign which is significant of myopathies should occur in amyotrophy one cannot say; it is mentioned, however, in the hope that an explanation may be forthcoming later.

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STATISTICS OF EPIDEMIC ENCEPHALITIS*

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THE MATERIAL

The following statistics are based on the records of cases of encephalitis which have been personally observed by numerous physicians throughout the country, both in their private and hospital practice. At first the attempt was made to gather the information from articles which have appeared in print, but even a cursory view of the literature soon convinces one of the futility of the task. Most of the articles are discursive; they deal with generalizations and contain descriptions of cases merely as illustrations of types or forms of the disease. The statistical facts which are desired are frequently incompletely recorded or altogether left out.[1] Valuable as many of the articles are they cannot furnish facts of sufficiently uniform value to make the basis of a comprehensive study.

In order to gather a sufficient number of cases to enable one to draw some definite conclusions a blank was sent to a great number of men who have had the opportunity of seeing many cases of encephalitis. The request was made to tabulate the age, sex, occupation, civil status, nativity, type or form of the disease, morbidity, occurrence of pregnancy, familial incidence, etc. About thirty neurologists generously responded to the request. Altogether the records of 864 cases have been collected, and these form the basis of this study. Practically every part of this country, as well as Montreal, Canada, is represented, though more than half of the records come from the city of New York.

The records of a number of hospitals have been obtained, and they are included in the statistics. Cases from clinics and dispensaries have been purposely left out, as many or most of them have previously been seen by consultants or were in hospitals whose records have been individually obtained. It is quite possible that "doubling up" has nevertheless occurred, as some cases have been seen by more than one consultant or were admitted to more than one hospital. This could not possibly be avoided particularly in the records obtained in New York City, but it is believed that the error is minimal and ought not to affect the results to any great extent.

The discrepancy in the various totals is due to the fact that in few instances the necessary data were left out in the records submitted.

AGE INCIDENCE

The accompanying table practically explains itself. The youngest cases on record were in infants of four weeks old. There were three such cases. One occurred in an infant of six weeks and one in one of seven weeks. The oldest case recorded is that of a

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[1] One author who read at least two papers before eminent medical societies and later published them, sent me incomplete records of two cases!

* Read before the Association for Research in Nervous and Mental Diseases, New York City, Dec. 29, 1920.

these patients with the extremely high mortality of influenza in pregnancy. The question of the influence of encephalitis on the offspring is of even greater interest, but is rather difficult to answer at present.

FAMILIAL INCIDENCE

In only five instances, a little over one half of one per cent, did the disease occur in two members of one family. There were two other instances, but both are doubtful and one of them occurred in a servant of the house. Considering the rarity of the instances one may well look upon them as accidental; and in view of the practical absence of contagion in hospital cases (two interns did contract the disease) one may consider the factor of direct transmission as almost negligible.

MORTALITY

Out of a total of 850 recorded, 178 patients died from the disease, giving a mortality of almost 21 per cent. (Generally speaking fatal terminations were most common in cases which began with high fever, those with very acute onset, in the so-called psychotic, toxic-delirious group, in patients with myoclonic symptoms and in those who showed severe meningitic involvement.) The remaining 672, or 79 per cent, do not, however, represent recoveries. It is true that the majority did recover more or less completely, but a great number showed the scars of the disease in the form of various sequellae. Parkinsonian residues, ocular manifestations, especially paralyses of accommodation, abnormal involuntary movements such as tics and tremors, epilepsies and psychic or even mental changes frequently were the heritage of the victims of epidemic encephalitis. In few instances the disease took on a chronic form: this was especially true of those with Parkinsonian residues and abnormal involuntary movements. Most of the patients, however, continued to improve even after many months of stationary chronicity.

CLINICAL FORMS AND ANATOMICAL LOCALIZATIONS

The great variety of clinical forms or types which were encountered make a uniform classification very difficult, if not impossible. The nomenclature varied with many observers, which added to the difficulty. Some divided, subdivided and classified their cases by creating types for every noticeable departure from the general picture, while others grouped them all under one or two heads. The anatomical substratum furnished justification for the former whereas clinical an detiological considerations guided the latter.

TABLE III.

Number	*Form*	*Per Cent*
303	Lethargic	35
129	Parkinsonian	15
89	Bulbo-pontine	10.3
66	Psychotic, Delirious	7.8
55	Toxic-Infectious Midbrain and Basal ganglia	6.3
44	Myoclonic	5.
43	Athetoid and Choreic	5
29	Radicular, Neuritic, Neuralgic	3.3
24	Meningitic	2.8
18	Hemiplegic	2.1
16	Myelitic, Spastic	1.8
11	Hemorrhagic	1.2
8	Convulsive	.9
7	Cerebellar, Ataxic	.8
5	Paraplegic	.6
4	Thalamic	.5
4	Catatonic	.5
3	Apoplectic	.34
2	Anterior poliomyelitic	
1	Posterior poliomyelitic	
1	Tabetic	
1	Pseudobulbar	
1	Optic Atrophy	

In many instances two or more classifications were noted for a given case. Most of the patients had lethargy added to the main clinical feature, but wherever a case was classified as lethargic plus some other form it was recorded under the special form. Some writers merely spoke of combined forms or lethargic and cranial nerve involvement. Others spoke of midbrain, basal ganglia, etc., and did not give clinical classifications. Obviously some of the headings could be combined, as one would have difficulty to decide whether, for example, a Parkinson syndrome should be recorded under basal ganglia or the clinical name. The psychotic, delirious, confusional group is very vague; some spoke of mental changes in general terms, others referred to a paretic form, while still others mentioned a Korsakoff picture. The very rare occurrence of pure thalamic syndromes is rather striking.

The term encephalitis has been altogether left out from the classification as too generic, since most of the cases were that, although meningo, myelo, radicular forms were not uncommon. The clinical classification ought really to be the guiding standard, as comparatively few cases came to autopsy, and most of the anatomical classifications were more or less inferential. However, the tabulated classifications reflect the opinions of the men whose cases are herein recorded. Table IV represents a composite statistical record of all the cases.

Grateful acknowledgement is hereby made to all who have generously contributed their case records to this study. Below is a list of the names of the contributors.

TABLE IV

Composite Statistics

Age	Sex	Occupation	Nativity	Civil Status	Mortality	Other Facts
Below 5 13 .... 1.5%	Males 522 or 60.5%	Practically every conceivable occupation	U. S. 387 or 45%	Married 491 or 56.9%	Died 178 or 21%	Pregnancies 22
6–10 37 .... 4%	Females 342 or 39.5%		Foreign 473 or 55%	Single 300 or 34.7%	Recovered, Improved or Chronic 672 or 79%	Familial Incidence. 5 instances of two in one family and two doubtful cases
11–20 136 ..... 15.7%				Not stated 73 or 8.4%		
21–30 222 ..... 25.7%			860	864	850	
31–40 215 ...... 25%						
41–50 140 ..... 16.2%						
51–60 72 .... 8.3%						
61–70 23 .... 2.8%						
Over 70 6 .... 0.7%						
864 100% Total						

Classification

Number	Form	Per cent	Number	Form	Per cent
303	Lethargic	35	16	Myelitic, Spastic	1.8
129	Parkinsonian	15	11	Hemorrhagic	1.2
89	Bulbo-pontine	10.3	8	Convulsive	.9
66	Psychotic, Delirious, Toxic-infectious	7.8	7	Cerebellar, Ataxic	.8
55	Midbrain and basal ganglia	6.3	5	Paraplegic	.6
44	Myoclonic	5	4	Thalamic	.5
43	Athetoid and Choreic	5	4	Catatonic	.5
29	Radicular, Neuritic, Neuralgic	3.3	3	Apoplectic	.34
24	Meningitic	2.8	2	Anterior poliomyelitic	
18	Hemiplegic	2.1	1	Posterior "	
			1	Tabetic	
			1	Pseudobulbar	
			1	Optic Atrophy	

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THE MORBID ANATOMY OF EPIDEMIC ENCEPHALITIS AS REGARDS THE ENDOCRINE SYSTEM*

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The histological examination of the glands of internal secretion in epidemic encephalitis was undertaken to ascertain whether any alterations were to be found, and if so, to determine, if possible, whether they could have any bearing on the course or manifestations of the disease.

A search through the autopsy records in the literature which has appeared up to the present time has failed to reveal any description of alterations in the internal secretory organs.

The material on which this paper is based consists of four cases. Each patient was observed prior to death, in the wards of the Presbyterian Hospital, New York City, and complete records of the clinical and neurological findings were kept. The autopsies were all made shortly after death. The material was fixed in Zencker's solution and the blocks embedded in paraffin. Sections were stained with hematoxylin and eosin.

CASE I. M. F., male, aged fifty-three. Three weeks previous to his admission he had pains in the eyes. Soon drowsiness and ptosis appeared and he suddenly became unable to speak or move his jaw, the mouth remaining partially open. On admission he was drowsy and extremely weak with ataxia of the lower extremities. Speech was almost unintelligible because of dysarthria. The deep reflexes were all active, but especially so on the right side. Plantar reflexes were normal, abdominals and cremasterics present. The eyes were directed forward and could not be moved voluntarily in any direction though they would follow an object moved laterally in front of them. The pupils reacted sluggishly to light. Slight papilledema was present in the left eye. The mouth was partially open and he could not move the jaw in any direction. Swallowing was almost impossible. The tongue could not be protruded beyond the lips, but showed a deviation to the right. The spinal fluid was under tension, contained nine lymphocytes, and gave a faintly positive globulin test. Death occurred from respiratory paralysis four weeks after the onset of the illness.

The microscopical findings on autopsy examination of the glands of internal secretion were as follows:

Thyroid.—The alveoli were of all sizes and filled with colloid. The lining epithelium was cuboidal. There was very little interstitial tissue and in places this appeared hyalinized or even colloid-like. There were a few localized collections of small cells in the interstitial tissue, with dense pyknotic nuclei. The vessels were greatly congested.

Adrenals.—The capsule was of average thickness. The cells of the cortex were well preserved and normal. The medulla showed moderate congestion but was otherwise unaltered.

Testicles were normal.

Pituitary.—Anterior lobe. The reticular arrangement was not very apparent as the cells were of large size and entirely filled the acini. The blood sinuses were dilated and congested throughout the entire lobe except for one localized area. The cells were predominantly basophilic. Those around the periphery were more intensely so, but even in the center they were more numerous than the other types. The cells were polygonal, with well defined outlines. The cytoplasm was finely granular. A few of the cells contained one or two vacuoles. These were usually in apposition with the nucleus. The nuclei were for the most part of large size, irregularly spherical, with a well defined reticulum. In the meshes of the reticulum were numerous chromatin granules. There were usually two or three large irregular chromatin masses and many smaller ones, the large ones being mainly toward the center of the nucleus while the smaller ones were principally at the periphery.

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* Presented at the First Annual Meeting of the Association for Research in Nervous and Mental Diseases, New York City, December 28, 1920.

In one portion the gland stood out in marked contrast to the appearance of the surrounding tissue. While the blood sinuses were congested in the other portions of the gland, here there were only a few small capillaries to be seen. The connective tissue stroma was swollen, edematous and fragmentary. The cells were of uniform staining and faintly basophilic. They were swollen and not well outlined, their margins being frayed and irregular. The cyotplasm was coarsely granular. The nuclei were frequently pyknotic, or occasionally swollen with large clear spaces betwen the chromatin network. In some instances there had been a rupture of the nucleolar membrane and a breaking up of its substance. Here and there were masses of protoplasm without nuclei or cell outlines. This entire area had the appearance of an early stage of coagulation necrosis from deficient blood supply.

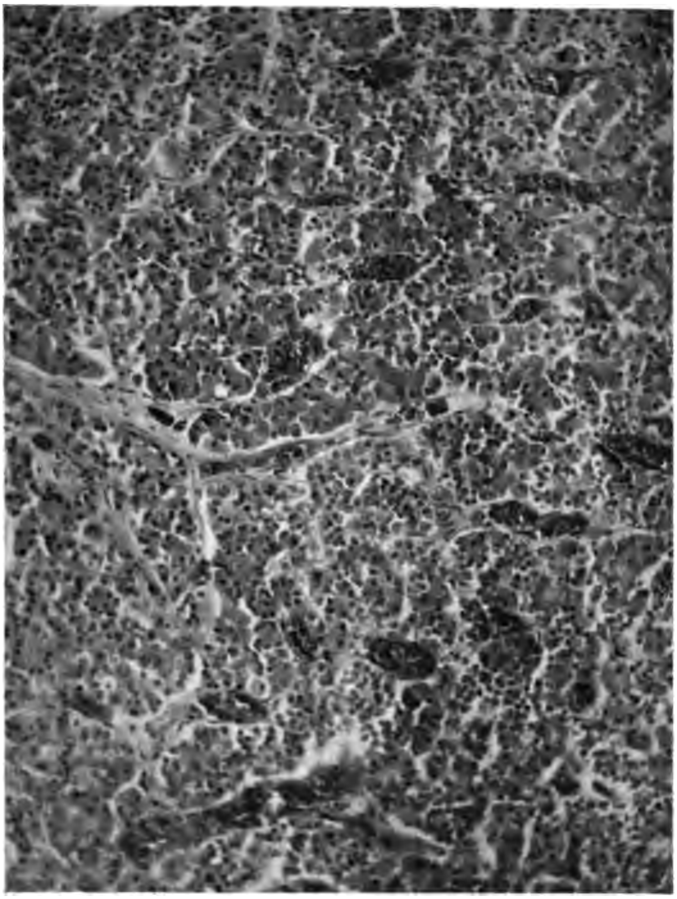

Fig. 1. Case 1. Pituitary Gland; Anterior Lobe. Large basophil cells and dilated blood sinuses.

Pars Intermedia. The cells were mainly faintly basophilic and arranged in the form of large vesicles which were filled with colloid.

Pars Nervosa. The neuroglial cells appeared normal. Some of the fibers contained a brownish pigment.

Case II.—A. M., male, aged fifty, whose chief complaints on admission were drowsiness, fever and twitchings of the muscles of the abdomen and legs. These symptoms had lasted for two weeks. At the onset he had rhinitis, headache and fever. Three days later there developed twitching of isolated muscles in all portions of his body. The generalized twitching continued for three days and then stopped to a great extent in the face, arms and chest, but continued in the abdomen and legs. Soon ptosis appeared with difficulty in swallowing and in urination. Weakness of the lower extremities with diminution of sensation on the legs and lower abdomen developed. The lethargy deepened to coma and death occurred five weeks after the appearance of the first symptoms. The spinal fluid was clear and colorless with normal pressure; it contained fourteen lymphocytes.

At autopsy the following findings were revealed:

Thyroid was small, rather soft and symmetrical. It appeared grossly normal. Microscopical examination showed the vesicles to be of average and about uniform size. They were filled with pinkish staining colloid. The cells were cuboidal and for the most part lined the acini in single rows. The blood vessels were not congested.

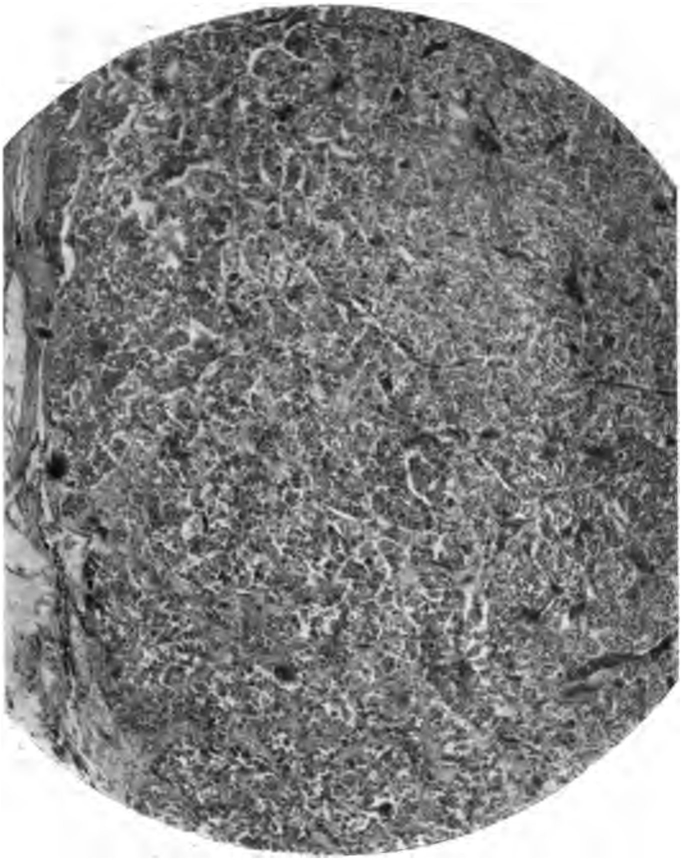

Fig. 2. Same as Figure 1. Low magnification showing normal cells and congested vessels at the right. At the left above, an avascular area with many swollen necrotic cells.

Pancreas.—The pancreas was congested but otherwise grossly normal. Microscopically it was normal except for a few large circular open spaces which may have contained fat. Some of them had a thin hyaline membrane lining.

Suprarenals were equal in size, small and flat. They contained considerable fat in the cortex with a rather broad inner zone and a small amount of white medulla. On microscopical examination the cortex was seen to contain a great deal of lipoid. At the junction of cortex and medulla were several large collections of round cells. In the reticular cortical zone was considerable lipochrome pigment. The medulla was broad, taking the chromaffin stain.

Testicles.—The left was soft and congested. The right, also soft and somewhat smaller than the left, was slightly adherent around the epididymis to the parietal layer of the tunica vaginalis.

Microscopically there was no spermatogenesis and practically all of the elements were absent, only the sustentacular cells and débris remaining in most of the tubules. The basement membranes were normal. The interstitial cells contained considerable brown pigment.

Pituitary.—Pars Anterior. The capsule was of normal thickness and throughout the gland there was a well-marked reticular arrangement of connective tissue. The blood sinuses were moderately dilated.

The cells had no definite arrangement but formed irregular groups. The great proportion of them were shrunken and in the center of the acini and not in apposition with their walls. The cells in the center were almost entirely eosinophilic, while those at the periphery were basophilic. The cells were of av-

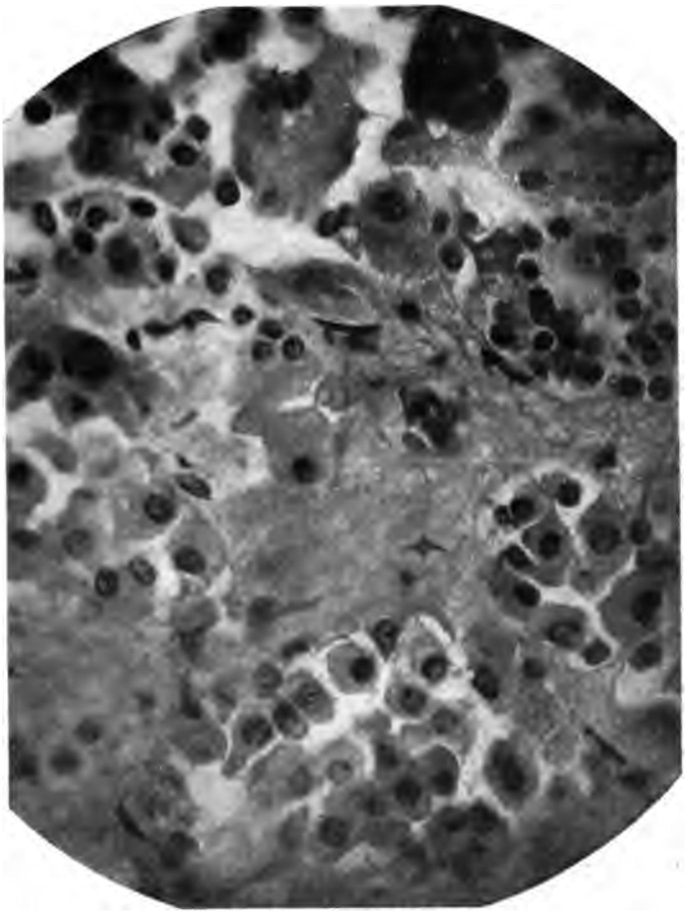

FIG. 3. SAME AS FIGURE 1. Higher magnification of avascular necrotic area.

erage size with well defined outlines and a single deeply stained nucleus in the center. Scattered throughout the entire anterior lobe were many large eosinophilic cells with a granular cytoplasm. These cells were more prevalent around the periphery, where they frequently were the only cells in an individual acinar space. Throughout the median portion of the gland they were rarely seen. The vascular sinuses were dilated. No hemorrhages or evidence of thrombosis were seen.

Case III.—N. F. Male, aged thirty-eight. The onset was sudden with an attack of dull, burning pain in the epigastrium. This was associated with nausea and constipation. After these symptoms had continued for two weeks, irregular twitching movements of the left arm commenced. At this time he had double vision and persistent hiccough. Drowsiness was noted about two weeks after the onset and this gradually deepened during a period of about ten days, when he became markedly stupid. On admission to the hospital he was in a semi-coma, sweating profusely, muttering incoherently and exhibiting jerky movements of the muscles of all portions of the body—face, neck, trunk and extremities. Bilateral ptosis, weakness of both external recti with incoordination of ocular movements, were present. The pupils reacted to light and on accommodation, but the right was larger than the left. Fundus examination revealed a slight blurring of the right disc. The extremities were spastic and the neck stiff. The deep reflexes were sluggish. The right knee jerk was obtained with difficulty; the left knee jerk and Achilles reflexes were absent. Urination and defecation were involuntary. The spinal fluid was clear and colorless but under slightly increased pressure, showing eighteen lymphocytes and a faintly positive globulin reaction. The lethargy deepened to coma, the temperature gradually mounted to 106, bronchopneumonia developed and death supervened after an illness of five weeks.

Pathological examination of the glands of internal secretion revealed:

Thyroid was of normal size and on section showed normal colloid material. The vesicles on microscopical examination appeared to be of average size. They were filled with a pink staining homogeneous colloid and lined by a single layer of cubical cells. There was no increase in the interstitial tissue and but moderate congestion of the blood vessels.

The Pancreas was grossly and microscopically normal.

The Suprarenals macroscopically presented a very yellow cortex and a deeply stained pigmented inner zone. On microscopical examination the glomerular zone was poor in fat, while the glomeruli of the vesicular zone were evacuolated. The whole gland was markedly congested.

Testes, on gross examination, apparently normal. On histological examination spermatogenesis appeared imperfect. Many pyknotic nuclear fragments were seen along with well preserved mitoses and apparently normal spermatis and spermatozoa.

Pituitary.—The connective tissue stroma was nowhere prominent but appeared norgestion of the vessels. The cells were mainly basophilic at the periphery while in the center they were distinctly acidophilic Neutrophilic cells were scattered irregularly throughout the entire section but were much in the minority, possibly one to eight or ten of the chromophilic cells. The eosinophilic cells were of average size. Their protoplasm was finely granular and did not contain vacuoles. The nuclei were generally spherical and deeply stained. They contained a chromatin network and granules, but were opaque. The basophilic cells were smaller than the oxyphilic. Their margins were less clear and more irregular. The cytoplasm was more coarsely granular and at times contained vacuoles. The nuclei were large and translucent with a chromatin network and granules. Two small vessels were thrombosed with what appeared to be platelet thrombi. Many of the cells around these vessels were necrotic. In some areas there was an accumulation of loosely granular cytoplasm without any nuclei or remnants, while in others the nuclei remained while the cytoplasm was disintegrated. Some of the nuclei appeared normal while others were fragmented and degenerated.

The pars intermedia contained many spaces which were filled with faintly basophilic colloid. The pars posterior was normal.

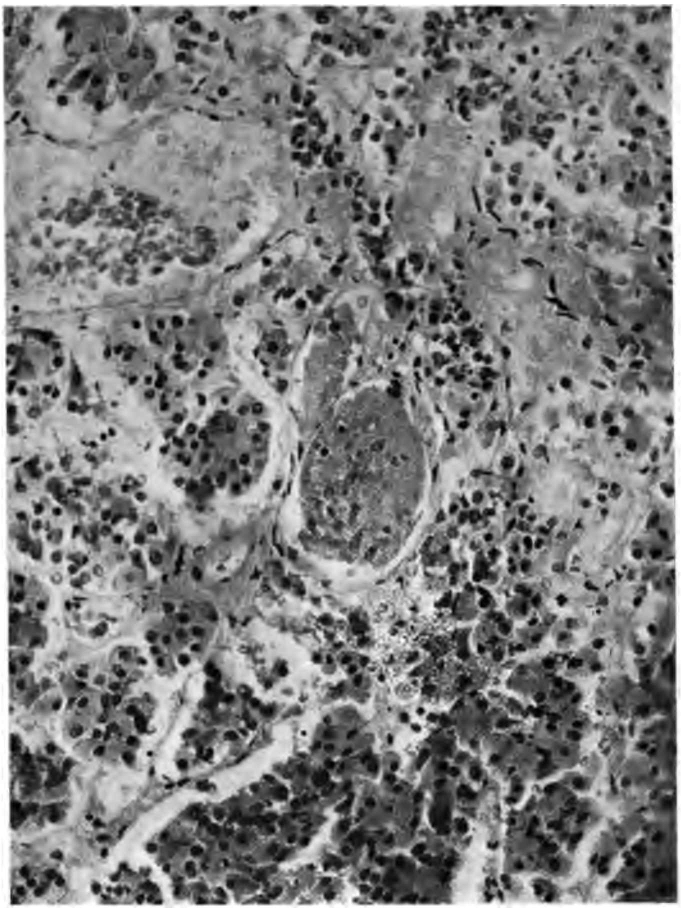

FIG. 4. CASE III. PITUITARY GLAND; ANTERIOR LOBE. Thrombosis of one of the small vessels, with necrotic area.

CASE IV.—Mrs. B. R., aged fifty-one, had been perfectly well until the onset of this illness. One morning she was found unconscious, rigid and frothing at the mouth. She regained consciousness in about an hour when she complained of weakness and tingling in the left arm and leg and marked drowsiness. She remained in bed in this state for a week. Then she got up and attempted to go around, but was very drowsy, frequently dropping off to sleep. She had a constant headache but no nausea or vomiting. This condition remained for two weeks when she was admitted to the hospital, complaining of headache, drowsiness and general debility. Examination at this time showed catatonic rigidity of the left arm and leg with increased deep reflexes and a Babinski on the left. Babinski on the right was absent. The muscle strength in the left arm and leg was perceptibly weaker than in the right. General sensory examination was not very satisfactory because of lethargy, but disclosed nothing abnormal. There was bilateral ptosis, more marked on the left; the pupils were markedly dilated, the left larger than the right. Reaction to light was sluggish. The eyegrounds were normal. There was marked weakness of the lower facial muscles on the left side. Blood pressure was 145-85. The spinal fluid was under much increased pressure, contained six lymphocytes and no globulin.

The lethargy gradually deepened into stupor and the left hemiplegia became complete. Urination was involuntary, and the patient died nine days after admission to the hospital, terminating an illness of about four weeks.

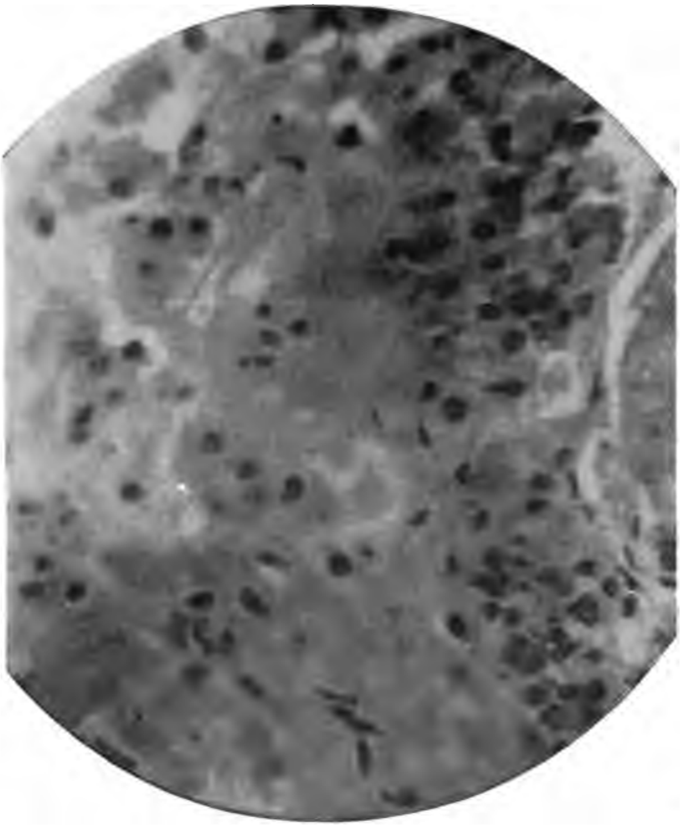

FIG. 5. SAME AS FIGURE 4. With high magnification of necrotic area.

The postmortem examination of the glands of internal secretion was as follows:

Thyroid was of normal size. On section there was possibly a slight decrease in the average colloid content. Microscopical examination showed the acini rather small and for the greater part lined with a single layer of epithelium. There was a slight increase in the interstitial tissue in which areas of hyalinization appeared.

Pancreas was of a reddish color with marked congestion of the blood vessels. The glandular tissue had a glossy digested appearance. On microscopical examination the pancreatic acini showed a decrease in size. The cytoplasm of the cells appeared vacuolated. Only a few of the islands of Langerhans were evident. Many were apparently replaced by fibrous tissue. There was a marked increase in the interstitial tissue replacing many glands and giving the tissue a fibrous appearance. In areas this connective tissue appeared necrotic. The blood vessels were sclerotic and many were obliterated.

Adrenals on gross examination appeared normal. On microscopic examination there was a thickening of the capsule and hyalinization. In the medulla the blood vessels were congested, otherwise nothing remarkable was seen.

Ovaries showed some old and some more

recent corpora lutea. Microscopically the ovary showed a few semi-replaced corpora lutea. Otherwise the entire ovary had been replaced with fibrous tissue.

Pituitary, grossly, showed marked congestion. Microscopically the reticular stroma was normal. The vascular spaces were dilated and many of them contained a homogeneous hyaline material. The staining reaction of the cells was mainly basophilic, but in the center the cells were mostly neutrophilic with a few oxyphilic scattered among them. Most of the basophilic cells were large and deeply stained. The cytoplasm was finely granular and rarely contained vacuoles. The margins were not well defined so the cells blended with each other. The nuclei were large, irregularly spherical or oval, and occasionally sausage-shaped or triangular. They contained a well marked chromatin network which supported dense granules. No thromboses or necrotic areas were seen. The juxtoneural epithelium and posterior lobe were missing.

CONCLUSIONS

There are some clinical manifestations that might be interpreted as endocrine disturbances. Among these are profuse sweating, weakness and low blood pressure, menstrual disturbances and impotence, and disorders of metabolism resulting in marked increase of weight. It seems probable that further study will show these disturbances to be dependent primarily on the lesions in the nervous system or the direct effect of the disease toxins on the individual internal secretory organs causing altered functions without distinct histological changes.

In the small amount of material available for this study, with the exception of the anterior lobe of the pituitary, there were no definite pathological alterations found in any of the glands of internal secretion which could have been the result of the terminal illness. In the pars anterior of the pituitary two kinds of alterations were observed: first, a predominance of basophilic cells, and second, areas of focal necrosis and capillary thromboses. The normal histological variations in the anterior lobe of the pituitary are marked and little understood. The significance of the preponderance of the various types of cells is largely hypothetical. There are two theories concerning the different cells. One is that the eosinophilic, basophilic and chromophobic cells are fundamentally different with different functions; and the other is that they are in reality one type of cell but of different staining properties in different stages of activity. The second view is, I believe, the one more generally held at present. Bell believes that the eosinophil cells predominate in the actively secreting gland. This is true of acromegaly where an eosinophil adenoma is usually found. He further states that the chromophobe cells are exhausted cells, while the basophil cells form a storage secretion.

To form any opinion of the state of secretory activity of the hypophyses described in this paper would be purely speculative, but it seems unlikely that there could be sufficient interference of function to cause the lethargy of epidemic encephalitis. The focal necroses and capillary thrombi are somewhat similar to the condition found in the liver in typhoid fever and in some cases of sepsis. Here, as in the liver, the collateral circulation is so extensive it is difficult to believe that capillary thrombi could impair the circulation sufficiently to induce necrosis. It seems more probable that both changes are produced by the circulating toxins. These necroses were not prominent in any of the specimens studied, and it is doubtful if they were sufficient to cause much alteration in the function of the gland.

So in conclusion we may state that, while the alterations described in the pituitary are definite, there is at present no evidence to show that they are of importance, or have any bearing upon the course or manifestations of the disease. In order that any definite conclusions can be reached concerning the changes in the internal secretory organs in epidemic encephalitis, many more cases must be studied.

ENCEPHALITIS LETHARGICA WITH A PROGRESSIVE PARALYSIS AGITANS SYNDROME

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The case presented to the conference is one illustrating the paralysis agitans type of encephalitis lethargica. The paralysis agitans syndrone is not uncommon in encephalitis lethargica, but the late onset of the signs and symptoms in this case, some of which did not appear until two years following the acute encephalitis, is quite uncommon and of especial interest.

R. F. (Case No. 30287), male, age thirty years, married, white, American; occupation, bicycle policeman.

HISTORY.—The present illness began in March, 1918, with quite a sudden onset. The patient was admitted to St. Lawrence Hospital, March 25, 1918. The hospital record, abstracts of which are below, shows that on the day of admission the patient complained of fatigue, great weakness and marked drowsiness; his face was flushed; temperature 103.2°, pulse 114, respiration 24. March 26, the drowsiness changed to a deep sleep with muttering delirium, retention of urine, patient catheterized; temperature 102°, pulse 100, respiration 20. March 27, irrational, slept greater part of day and night, body apparently stiff, temperature 102°, pulse 100, respiration 20. March 28, irrational, in a deep sleep, vomited, retention of urine, temperature 100.8°, pulse 100, respiration 20. March 29, irrational, carphologia, abdomen greatly distended, temperature 100°, pulse 96, respiration 20. March 30 to April 2, condition unchanged. April 3, report of blood examination: Widal reaction does not show. April 4, sleeps for long periods, but awakens brighter and is rational, temperature 99.8°, pulse 94, respiration 18. April 5 to 12, drowsy for shorter periods and feels comfortable. April 17, drowsy, out on roof six hours, temperature 99°, pulse 92, respiration 18. April 21, out of bed, very listless and dull, temperature 98.8°, pulse 86, respiration 18. Eye examination: Pupils react normally to light, distance vision apparently normal, paresis of accommodation. April 27, out on roof greater part of day, listless and dull. Temperature 98.4°, pulse 90, respiration 18. May 4, 1918, discharged to home.

At arrival at home his only complaint was of a general weakness and an inability to walk more than a few paces without great fatigue. June, 1918, he noticed for the first time that objects appeared double to him. This condition of diplopia lasted until March, 1919. June, 1918, about one week following the onset of the diplopia, he began to have a slight tremor of the right foot and leg. His body strength gradually increased and he walked with little or no difficulty. October 1, 1918, following the examination by the Police Surgeon, he resumed the duties of a policeman. He got along well except that he became more easily fatigued than he did when on duty prior to his illness. He rode a bicycle well. In March, 1920, he began to have a tremor of the right hand and arm, and in April, 1920, a tremor of the left foot and leg. The gait became progressively more unsteady; it was noticed that upon turning quickly, he had great difficulty in preventing himself from falling. When walking down stairs he was prone to go too fast, and in August, 1920, he fell heavily down stairs. He became emotionally unstable, was more irritable, impatient and readily cried upon slight provocation. It became necessary to cease bicycle riding and in August, 1920, he found it necessary to report on "sick leave" to the Police Surgeon. He has not worked since because of a progressive increase in the tremor, a gradual increased difficulty in walking, and a feeling of systemic weakness.

October 31, 1920, he was admitted to St. Luke's Hospital. He remained in the hospital two weeks and then was discharged to his home. During this period of observation, the temperature ranged from 97° to 101°, pulse rate 78 to 96, and respiration was normal. The blood examination showed red blood cells 5,300,000, Hemoglobin 107 per cent. White blood cells 11,500, the polynuclears 82 per cent, and lymphocytes 18 per cent. Wassermann reaction, negative. The spinal fluid examination showed a clear, colorless fluid under slightly increased pressure. Cells, 0. Wassermann reaction, negative. Colloidal gold 1111.5210.5000. Urinalysis was negative.

NEUROLOGICAL EXAMINATION

Facies ironed out, of unemotional type, right palpebral fissure less than left, pupils normal. There is a left facial weakness, motion is possible but decidedly rigid, the right arm and leg show a weakness of motor power. Abdominals, left greater than right. Knee jerks and ankle jerks, right greater than left. Chaddock on right is questionably positive. No other extensor reflexes. There is a coarse oscillatory tremor of the right arm and leg and left leg, which ceases on intention, a slight rigidity of movements of the right side of the body and a lack of coordinate movements.

November 13, 1920, discharged from St. Luke's Hospital to home.

The patient was admitted to Vanderbilt Clinic, November 29, 1920, with the following complaint: Tremor of head, right arm and leg, left leg, and an unsteadiness in walking, particularly noticed upon changing direction or turning about.

Injuries and Disease.—None except at twenty years of age he had gonorrhea, which lasted four months. Syphilis denied.

Habits and General Physiological Status.—Very moderate tea and coffee drinker, rarely indulging in alcoholic drinks. Smokes about 15 cigarettes a day, and at the present time gets great comfort from them because of a cessation in his tremor after smoking half a cigarette. Sleeps and eats well and his wife has noticed that the tremor ceases during sleep. Thirst is increased. Weight about normal, 180 pounds. No urinary troubles. Bowels regular. No sexual habit since June, 1918. Potency impaired since June, 1918.

Family History.—Patient has been married six years and wife has never become pregnant. Father is dead, cause unknown. Mother died at childbirth. He has one brother alive and well.

VOLUNTARY MOTOR SYSTEM.—*Abnormal Attitudes and Deformities.*—Stands with a slight forward stoop, head and neck held stiffly, right arm held in semi-flexed position, right hand assumes the accoucheur position, although not constantly so. The palpebral fissures are unequal, left greater than right. There is a lower facial flattening and the impression of a stare is given.

Gait.—Walks slowly with a tendency to propulsion, the normal heel-toe gait is impaired, he strikes the floor with the heel and ball of foot more nearly at the same time, right arm held semi-flexed, with impaired associated movements. Upon walking backward, there is a tendency to retropulsion. He stands on toes normally, but is very unsteady when he stands on heels. Walks sideways and runs normally. Fournier's tests are positive.

Coordination.—Patient is right-handed. Equilibratory: Stands normally with feet together, eyes open. Romberg is absent. Stands normally on left foot, but has slight unsteadiness when he stands on right foot. Non-equilibratory: Finger-to-nose, finger-to-finger, each-finger-to-thumb, all these tests done with eyes open and closed are performed normally, but accompanied with a slight tremor in the fingers and hands and the right forearm. Pointing and pass-pointing, left and right, are done normally. Heel-to-knee, and along shin, toe-to-object above recumbent patient, both tests done with eyes open and closed, are performed normally, but accompanied with a slight tremor on the right side. Adiadochokinesis, absent on left but present on right. No dysmetria, right or left.

Skilled acts.—Speech has a low, monotonous tone, there is no apraxia or aphasia. The handwriting shows a fine tremor.

Abnormal Involuntary Movements.—There is a coarse, slow, rhythmical, non-intention tremor, which varies in its amplitude, but is of regular force. The tremor involves the head, right arm and leg, and left leg, and is decreased by action and volitional effort, and increased in repose and emotion.

Reflexes.—(a) Deep: left upper extremity, are active; right upper extremity, are very active; lower extremities, suprapatellar, left and right, very active, patellar, left and right, very active, patellar, left and right, very active, patello-adductor, left and right, absent; tibio-adductor (periosteal) left and right, absent; tibio-adductor, right to left and left to right, absent; cuboido-digital (dorsal flexion), left and right, active; Achilles, left and right, active; ankle clonus, left and right, absent. (b) Superficial; ciliospinal, left and right, present; abdominals, left and right, active; cremasteric, left and right, active; plantar flexion, left and right, slight; Babinski, Chaddock, Oppenheim, Gordon, Schäfer, left and right, are all absent.

Muscle Strength.—Unable to rise from recumbent position to sitting, and if the movement is done slowly, he is unable to rise from sitting position to standing. If a sudden or quick attempt is made to rise from a sitting position to standing, the act is performed normally. Unopposed movements are not limited in extent. Opposed movements of the head, right upper and lower extremities, and left lower extremity, show a degree of strength slightly less than normal.

Muscle Status.—The volume and contour show no atrophies or hypertrophies. Consistency is normal, tone, as tested by resistance to passive movement and extensibility of joints, shows a hypertonus of the right upper and lower extremities, left lower extremity and post-cervical group of muscles. Irritability and myoidema are absent.

Abnormal Associated Movements.—The Hoffman sign and thumb sign of Klippel-Weil, left and right, are doubtfully present. The sign of Grasset-Bychowski is absent on the left and right.

GENERAL SENSORY.—Touch and touch acuity, pain (superficial and deep), temperature (cool and warm), vibratory, muscle tendon, and stereognosis are normal.

CRANIAL NERVES. *Olfactory.*—Smell (subjective) normal.

Optic, Oculomotor, Trochlear, Abducens Nerves and Optic Apparatus.—Vision, left and right, acuity and fields are normal. Fundi are normal. Pupils, left equals right, 3 mm. in diameter. Shape, the left is irregular; the right is circular, position is normal. No hippus present. The reaction to light, direct, left and right, is prompt but not of great amplitude; consensual, both right to left and left to right, is normal. Reaction to convergence, left and right, is normal, and there is no Argyll Robertson pupil. Position of the eyes is normal and there is no diplopia. There is no limitation of voluntary movement. A nystagmoid movement, lateral in direction and of small amplitude, is present to left and right. Palpebral fissures: It is a question whether the inequality may not be due to a widening of the left or an increased rigidity causing a narrowing of the right. No Dalrymple, von Graefe or Stellwag. No abnormalities of iris, lens, media, cornea or conjunctiva.

Acoustic Nerve and Ear.—(a) Cochlear: Hearing, subjective and objective, is normal. (b) Vestibular: Caloric, rotary and Hitzig tests not made.

Trigeminal Nerve and Mouth.—Masseter muscle of normal strength. Corneal reflex, left and right, is normal. Teeth in excellent condition.

Facial Nerve and Face.—Taste, subjective, is normal. The movements of the left upper and lower facial muscles are normal; there is an impaired motion of the right lower facial muscles accompanied with a slight tremor, and a slower action and lessened wrinkling on the right side of the forehead. The labial articulation is normal, but the associated movements of the right side of the face are impaired. No change in the vasomotor or secretory status.

Glossopharyngeus and Vagus Nerves.—Taste is normal, position of uvula central and the movements of the palate during phonation are normal. The voice is low and and of an unmodulated tone. There is no difficulty in swallowing.

Spinal Accessory Nerve.—Is normal.

Hypoglossus Nerve and Tongue.—The tongue is protruded in the midline, and the movements in all directions are normal.

MENTAL STATUS.—The patient is intelligent, with good attention and memory; but he is quite emotional, crying easily. There are no phobias or compulsions, hallucinations or delusions.

SYSTEMIC. *Tegumentary.*—Hair well-distributed over abdomen and extremities, the beard is even and fairly heavy. There are no areas of pigmentation and the skin is smooth, moist, warm and with no eruptions. He perspires freely over the entire body. There is no edema. Sergent's line is present.

Glandular.—Thyroid seems normal in size and the tonsils are not hypertrophied. No nodes palpable in anterior and posterior cervical, axillary, epitrochlear, or inguinal regions.

Cardio-Vascular.—Pulse rate is 80, it is regular and easily compressible. Blood pressure, systolic, 140 mm. and diastolic 110 mm. Heart sounds are regular, but of poor quality and force. No murmurs are heard.

Respiratory, Gastro-Intestinal, and Genito-Urinary Systems are negative.

LABORATORY EXAMINATION.—December 1, 1920, Wassermann reaction of the blood, negative.

SUMMARY AND CONCLUSION

A young man in good health, suddenly became acutely ill. The symptoms of onset were fever, great weakness, fatigue and lethargy. In a few days there was rigidity of the body, followed by a muttering delirium, carphologia and a loss of the expulsive power of the bladder and rectum. The Widal reaction did not show. The fever gradually lowered and the patient was out of bed in about five weeks, but was quite listless and dull. About three months following the onset of the illness, a diplopia and a tremor of the right foot were first noticed. A few months later work was resumed, the weight and strength increased, but the robust health possessed prior to the illness was never regained. Eighteen months later a tremor appeared in the right hand and a few weeks later in the left foot. The gait became progressively more unsteady and there was an increased emotional instability. There was an inability to continue work and he entered the hospital for observation and treatment. The Wassermann reaction of the blood and spinal fluid was negative. The spinal fluid was under slight pressure, but showed no cells. Other blood examinations and the urinalysis were negative. The symptoms have progressively increased and upon admission to Vanderbilt Clinic, there was shown a well-developed Parkinsonian syndrome. The initial symptoms, particularly the lethargy and the rigidity, are interpreted as clinical manifestations of an acute inflammatory process with a severe toxemia involving the basal ganglia. The diplopia was undoubtedly of midbrain origin. Later the appearance of a non-intention tremor involving the right foot and leg, right hand and arm, left foot and leg, accompanied with an increased muscle tone of these parts, and loss of associated movements of the right arm and right side of the face, showed further involvement of the basal ganglia, and, according to the view of J. Ramsay Hunt, these symptoms are due to a destruction of the large cells of the globus pallidus system. The case is undoubtedly one of acute encephalitis lethargica with an unusually long interval before the appearance of the paralysis agitans type of sequel.

DISCUSSION

Dr. McKendree asked whether the patient had been entirely well during the two years after his attack.

Dr. Blakeslee replied that he had not been entirely well, fatiguing much more quickly than before his illness. But he had been able to ride his bicycle very well.

Dr. Lehrman asked whether the corneal and pharyngeal reflexes were present, and was told that they were.

During the reading of the history of the case, the patient had been given a cigarette.

Dr. McKendree remarked that the tremor was absent in the hands while the patient smoked, but not in the legs. This might have been attributable to the sedative action of the cigarette or might suggest a psychic condition.

Dr. Polon said he was interested in the group of symptoms Dr. Blakeslee had mentioned—excessive sweating and Von Graefe. From the element of vagotonia he would not be surprised if the patient had a very slow pulse. No mention of the pulse rate had been made.

Dr. Polon also emphasized the importance of the patient's ability to get up quickly and inability to get up slowly and believed this should be borne in mind for therapeutic reasons. One of his patients had been very much concerned by the slowness of his movements and was greatly relieved when he learned that he could do things quickly that were impossible for him to do slowly. Another case he knew of, with paralysis agitans symptoms after encephalitis, could not walk, but could run so quickly that it was impossible for him to stop; someone had to stop him.

Dr. Tilney said that there must be some explanation of the long gaps between the acute state of encephalitis and the development of new symptoms. It was more than a recrudescence, as it appeared to be a development of an entirely new clinical picture. He supposed it was fair to start with some working hypothesis as to why this may be the case. One possibility that suggested itself was that the organism becomes walled off somewhere in the nervous system so that there was an arrest, which later breaks out again, and organisms being liberated in the circulation, start a new focus giving an entirely new picture in the disease.

Another possibility is suggested by the nature of the infection. We feel that there is much evidence to-day, although disputed, that the organism gains entrance to the nervous system from the nasopharynx. The work of Strauss and Loewe points in this direction. If this is so, we do not know how long the organism may live in the nasopharynx, and living there for some time, it may start a secondary invasion.

This case was particularly interesting to Dr. Tilney because of a patient Dr. Riley and he have been watching at the Roosevelt Hospital for four months. They saw him during his lethargy which persisted for a number of weeks. Then he seemed to be getting well. He had a relapse, however, from which he is just beginning to clear. In the latter part of his relapse Dr. Strauss found the organism he has already described in the nasal washings and was able to transmit it to a rabbit, causing death due to encephalitis. So that we have evidence here of the long persistence of the organism in the nasal secretions.

Dr. Tilney was glad that Dr. Polon had emphasized the difference in the patient's ability to rise slowly and to rise quickly. These were important things to remember in treating such cases which had been neglected up to the present time.

Dr. Casamajor stated that paralysis agitans brings out some interesting points. The retropulsion in this patient was quite marked. Another interesting point was the way he sat down on the chair. He came down with a thud, striking one shoulder on the edge of the chair. How different that was from the way a tabetic flops down! Had a tabetic struck the chair as this patient did, he would have fallen on the floor. The patient resembled cases of manganese poisoning whose only symptom was their inability to sit down slowly. These paralysis agitans patients often find out for themselves the point brought out by Dr. Polon, about their ability to do things quickly. That may be a special type of paralysis agitans difficulty, or it may be that the patient has learned the fact that he gets along well when he goes quickly, or has someone to pace him, but otherwise "runs down." Dr. Tilney described this condition some years ago and called it "metadromal progression."

Dr. Osnato thought that there was evidence of a recent flare-up in the epidemic of encephalitis. For some time he had seen no new cases, until this week when he had seen three. He wondered whether other physicians had met with the same experience.

POST-ENCEPHALITIC INVOLUNTARY MOVEMENT

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CASE I. This patient (Case History No. 35444), a Russian Jewess, twenty-five years of age, married, came to the Vanderbilt Clinic on December 23, 1920, complaining of an involuntary, jerky, movement of the left lower extremity, manifesting itself when sitting or reclining, and to a less degree when standing; it disappears in sleep and in walking and whenever the affected limb is actively engaged, as in a rotary or kicking-out motion. It also is absent when the body is supported by the left limb alone. Apparently the motion causes but little disability or discomfort.

The affection first manifested itself in the middle of July, 1920, three months after recovery from what the patient calls the "sleeping sickness," and two months after having given uneventful birth to a normal child.

The onset was gradual. The first symptom was a slight adduction twitch of the left thigh, occurring regularly every few minutes. As time went on the twitching movement became more and more frequent, and more and more extensive in excursion, reaching its greatest intensity by the end of October, 1920, about three months after the onset, and remaining the same since then.

The history of the above mentioned "sleeping sickness" is as follows: Quite well until, about the end of January, 1920, when she was gravid in the sixth month, she became subject to insidiously increasing headache, fatiguability and somnolence. Her efforts to fight them off by persisting at work were in vain. By the middle of February, 1920, she took to bed with fever. Her condition was then diagnosed as "sleeping sickness."

Within a few days the somnolence became much more marked; she slept most of the time, and had to be awakened for food and drink, which she took without resistance. In the intervals between the lethargy she showed marked confusion; did not recognize her relatives; hallucinated; saw a child thrown from the roof of the house, insisted that she had already given birth, and looked for the newly-born under the pillows; was quite agitated; was kept in bed with difficulty, attempted bodily harm to her attendant, tore up her clothes, and fumbled with her blankets. Her eyes became staring and wild, the speech slurring, indistinct and monotonous, and her face drawn over to the right. When not in this delirious state she would lie in a deep sleep, perspiring profusely.

At first she had to be catheterized, but after a few days urination became normal. At no time was there urinary or fecal incontinence. An unreliable history of diplopia and possibly left internal strabismus was obtained. Fever was present only for a few days and was not high at any time. There were no signs of threatened abortion.

After being in this state for about two weeks she was lumbar punctured, and within twelve hours showed improvement which steadily and slowly progressed. First her mental condition cleared, then the speech. and finally the somnolence and facial asymmetry receded. By the middle of April after being in bed for about eight weeks, she gradually returned to her usual occupation. Except for some irritability and diminished vivacity, according to her family, there were no after effects of the illness.

On May 11, 1920, about one month after recovery, she gave normal birth to a normal child. Puerperium and early months of lactation were uneventful. She worked about the house, nursed the child, was gaining in weight, her sensorium was clear, sleep, digestive and excretory functions were normal. There was no evidence of weakness, tremor, or any abnormal movement until the middle of July, 1920, when she first observed a slight twitch in the left lower limb and which eventually became the movement that constitutes her present complaint. The change from a placid disposition to that of an explosive kind, plus some immobility of the face with an asymmetry of the ocular apertures, are symptoms which her family believe date from the time of the "sleeping sickness."

Heredity, development, habits, menstruation are negative. She has not menstruated since August, 1919. She is still nursing her eight months old infant. Besides the mentioned "sleeping sickness" she had measles in childhood.

A native of Russia, she emigrated to the United States nine years ago. She married five years ago, and gave birth to two normal children who are alive and well at the ages of three years and eight months respectively. The younger one she carried while she was ill with the sleeping sickness. There were no miscarriages.

PHYSICAL EXAMINATION

VOLUNTARY MOTOR SYSTEM.—Abnormal attitudes and deformities are not observable except for slight facial asymmetry (weakness of left) and some stolidity of facial expression. Gait is quite normal in walking and running in all its phases, pelvic, thigh, leg and foot. No abnormal involuntary movement is manifest during the tests. Fournier's tests are negative. *Coordination:* Equilibratory.—Standing with feet together eyes open and closed, no Romberg. Nonequilibratory.—Finger-to-nose, finger-to-finger, toe-to-object, heel-to-knee and dysmetria tests are negative. There is perhaps a slight adiadochokinesis of left hand. Skilled acts.—There is no dyspraxia or apraxia. The speech appears to be somewhat slurred. There is no aphasia.

Abnormal Involuntary Movements.—There is an abnormal involuntary movement limited exclusively to the left lower limb, which varies somewhat depending upon the posture. When sitting, there is an adduction of the thigh, followed by an extension of the leg, and eversion of the foot followed by a dorsal flexion of the toes. The latter is suggestive of an athetoid movement. When standing there are dorsal flexion of the toes, flexion of the knee and thigh, causing trunk to bend forward and to the left. When lying down there are flexion of thigh raising the limb about one inch from bed, flexion of knee with visible contraction of quadriceps tendon and slight adduction of thigh. Occasionally there is also dorsal flexion of all toes suggestive of athetosis, or only dorsal flexion of the big toe.

The frequency of this movement is once every four seconds. Its duration is one second. Volitional inhibition does not affect the interval but somewhat modifies the extent of the movement, and the patient experiences an uncomfortable feeling. Manipulation of the limb, stroking or pricking it, intensifies the athetoid character of the toe movement.

These rythmic muscular contractions are those of synergic groups, causing considerable displacement of the limb. While bearing some resemblance to chorea they are less rapid than those ordinarily observed in chorea and have no explosive character. They are constant, involving always the same muscles. This imparts to the movement a psychic content and suggests a tic. The contraction commences in the adductors of the thigh, perhaps sartorius, followed by an extension of the leg, and finally by the foot and toe movement. These movements do not appear to occur simultaneously, but manifest themselves in the order just mentioned, starting proximally and terminating distally. The slowness, labored character and sinuosity of the muscle contractions plus the displacement of the limb caused by them, suggestive on the one hand of chorea, and on the other of athetosis, would fittingly be designated by the term choreo-athetosis. There is no tremor.

All deep and superficial reflexes are quite normal or perhaps a little hyperactive, but there is no appreciable difference between the two sides. There is no ankle clonus, no

Babinski, no loss of abdominal reflexes, etc.

Tests of muscle strength, passive and active, reveal no abnormality. The muscle status of both sides is equal and normal. The left calf equally with the right measures 28 cm. about its fullest portion. There is no rigidity.

Touch, pain, temperature, vibratory, pressure and muscle joint tendon sense, as well as stereognosis and barognosis, are normal.

Cranial Nerves.—The olfactory nerve is normal. The Optic.—The acuity and fields of vision are normal, so are the fundi. Pupils are equal, 3 mm. in size, regular in shape, central position, react to light directly and consensually, normally. The pupillary reaction to convergence is sluggish. There is no strabismus. On looking to the right and upward diplopia is elicited. There is a bilateral slight enophthalmus. There is a Möbius sign on the left side. There is a rapid rhythmical nystagmoid movement on looking to the left. These are not constant and probably depend upon a residual weakness of the ocular muscles. There is an inequality of the palpebral fissures, the right being larger. The acoustic nerve and ear reveal nothing abnormal, nor does the fifth nerve. The left face appears flattened and drooping. The left ocular aperture is smaller than the right. This inequality disappears in active innervation of the face and in emotion. There is no change in the taste function. The vasomotor and secretory status are negative. No abnormalities are noted in the ninth and tenth, eleventh and twelfth nerves.

Mental Status.—There is no evidence whatsoever of confusion. Her intelligence is good corresponding to her educational level. Emotion, attention and memory are normal.

Systemic.—Tegumentary. The only thing of note is a diffuse hypotrichosis. There is a complete absence of hair in the axillae.

Glandular.—The thyroid gland is not palpable. *Cardio-vascular.*—The pulse is regular, well-sustained, frequency 72, blood-pressure is 105/70. The heart is negative.

Lungs, abdomen, genito-urinary and skeletal systems are negative.

No laboratory findings were available.

SUMMARY OF HISTORY

1. Encephalitis, February to April, 1920, with marked psychotic and few paralytic symptoms in a six months gravid woman, followed by complete recovery.
2. Apparently normal health for the succeeding three months, during which she gave normal birth to a normal child, and is still nursing it.
3. Sudden appearance of a choreo-athetotic involuntary movement, without any other signs of disturbed function, three months after recovery. This movement persists to date. It causes but little discomfort and does not interfere with patient's usual course of living, or with her nursing the child.

The outstanding features of the physical examination are as follows :

1. The choreo-athetotic movement of the left limb characterized by (a) contraction of synergic groups of muscles; (b) regularity and rhythmicity—the frequency being about once every four seconds and duration one second; (c) slowness, sinuosity and labored character; (d) the comparative non-responsiveness to voluntary inhibition, and (e) its complete disappearance when limb is engaged in the execution of active motion.
2. Some residual palsy of the ocular muscles, manifested by (a) slight limitation of pupillary accommodation reaction; (b) diplopia on looking to the right and upward; (c) left möbius; (d) nystagmoid movements, and (e) right palpebral fissure larger than the left.
3. Some stolidity of facial expression and a very slight weakness of left face of supernuclear type.
4. Absence of pyramidal tract involvement.
5. Absence of sensory disturbance.
6. Absence of lower motor neurone involvement. No fibrillation, no spinal myoclonus, no atrophy, no weakness.

7. Hypotrichosis, hypotension, hypothyroidism.

DIAGNOSIS

The history of encephalitis during the epidemic, three months before onset of the present complaint, the presence of residual palsies of ocular muscles, the involuntary movements of a choreo-athetotic nature, all point to a post-encephalitic condition.

Because of the stereotypy of the movement, hysteria suggests itself, but the absence of hysterical make-up, hysterical sensory signs, and the presence of signs of organicity permit of an exclusion of a functional disturbance.

The lapse of two months between the puerperium and the appearance of the movement speaks strongly against either chorea gravidarum or puerperal chorea.

Localization.—One is thus dealing with an organic condition which manifests itself by positive as well as negative symptoms, using these terms as H. Jackson intended them to connote. The positive symptom in this case is the choreo-athetosis, which may be regarded either as an expression of an irritative lesion, or as the result of release of a center, due to a destructive lesion of a higher center.

On the ground that the involuntary movement is the result of contraction of synergic groups of muscles, spinal localization may be excluded. Groups of muscles are not represented in the cord, or only negligibly so. Furthermore, the usual spinal irritation phenomenon, such as fibrillation or myoclonus of portions of a muscle or of a single muscle is lacking in this case. The lesion is evidently located in the cerebrum.

The cortex is not likely to be the seat of the lesion, either as a parenchymatous change, or that due to a meningeal irritation. Were it so, convulsions Jacksonian or generalized, or other cortical focal phenomena would result. These are lacking in this syndrome. The absence of pyramidal tract involvement excludes the pre-Rolandic area. The intactness of mentality speaks against the involvement of the cortical association pathways.

The same considerations apply to the centrum ovale. The absence of sensory disturbances point against a thalamic involvement. There are no cerebellar, medullary or pontine signs.

The mesencephalon offers no likelihood of localization. Were its tegmental portion involved in the sensory pathways or in the nucleus ruber, or in the substantia nigra, or in the nuclei of the third and fourth, and the mesencephalic root of the fifth nerves, we would have sensory disturbances, rigidity, tremor, pronounced ocular palsy, and some difficulty in mastication. All these are lacking. The crusta are apparently free, as evidenced by the absent pyramidal signs.

One is thus limited in localization of the movement to the corpus striatum. According to Morat, "The gray masses of the corpus striatum contain within themselves associations of sensory and motor elements, which transmit the impulse in the desired manner, so as to secure the performance by the limbs of the movements by means of which progression is effected; and, it may be added, its continuance when once commenced. These movements are automatic; or to put it differently, involuntary. They are produced by the arrival in the system which is adapted to their performance of an impulse which initiates them. They cease when another impulse arrests the preceding one and brings the automatic system back to a state of repose; they continue during the interval between these two impulsions. The initiation or its cessation is alone voluntary, and therefore the impulse which produces it comes from the cortex, or must have passed through it." In this case the mechanism whereby a cessation of an automatic movement could be brought about is impaired, whereas the mechanism concerned in the initiation of the automatic movement is intact. Hence, the patient is capable of a voluntary execution of this motion, but is not able to inhibit it.

The question whether this affliction represents a continuance of the original encephalthat I have seen the element has been that of extension. I have not as yet seen any real flexion; at least, they start with extension, they may end up with flexion.

I read an article by Wilson, who starts off with a reference to the work of Sherrington in the study of apes. Wilson considers these athetoses as a part of a decerebrate picture and he thinks that in the main the causation of the thing lies in the red nucleus, because in these decerebrate apes tonic fits were caused by stimulation of the red nucleus. Wilson states that there is a definite pattern, a definite arrangement of cells in the red nucleus. Certainly the cortex has a definite pattern; it has its cells arranged in different layers, so that stimulation of certain parts of the cortex causes contractions of a very definite group of muscles. The cerebellum, we know, has also a pattern. I would not be surprised if we were to find a pattern in the lenticular and caudate nuclei. Wilson believes that in these so-called tonic fits of the decerebrate animals the thing is reflex; the stimulus arises first on the outside. It does not travel up the posterior column because section of the posterior column does not stop the tonic fits. Wilson believes that the stimulus is carried up to the cerebellum first and then to the red nucleus.

I think the interesting thing would be to find out where we are going to get the time element. The cortex gives to all sensations a time element. I think it is doubtful if the cerebellum has a real time element. If Gaskell's concept of the primary function of the basal ganglia is correct, we must certainly give to the corpus striatum and the basal ganglia a time sense.

Dr. Tilney's work is going to be a great help in finding out what this pattern is and whether we can compare it to a cortical pattern. I think it is only by such work as Dr. Tilney contemplates that we can answer that question.

So far as I know, there has been no literature upon the subject based upon real pathological studies, except this article of Wilson's which is rather far removed from these particular cases.

Dr. Polon. These two cases and the cases reported by Archambault, have all had severe psychotic symptoms at the onset. I do not remember one in whom there were not psychotic symptoms in the initial onset of the encephalitic process.

I wonder if a movement which the patient can start but cannot stop can be attributed to the cortex. The cortex is only capable of initiating the movement. The automatic part is most likely taken care by the corpus striatum. Automatic movements have been described by other writers in relation to the corpus striatum.

I should mention that in the recent contribution of Marie and Levy, they have found it very difficult to classify these post-encephalatic movements in terms of the old terminology. But I suppose, for want of a better name, choreo-athetosis will have to be accepted until further elucidation.

Dr. Osnato. I would like to ask Dr. Polon what he tells these patients as far as prognosis is concerned.

Dr. Polon. The experience of most men is that these cases go from bad to worse and that the prognosis is decidedly unfavorable. The first case I presented has shown no change for three months. She has gained considerably in weight, but since her nutrition has been built up there has been not the slightest change in the movement. I have given a guarded prognosis. I have wondered what re-education movements would do. It is very likely that the cortical impulse which has to do with this movement is not completely cut off, and that by re-education some improvement might be brought about.

Dr. Osnato. In an article I read recently, the author stated that the majority of these cases get well and a great many of them are greatly improved.

EPIDEMIC ENCEPHALITIS WITH UNUSUAL SEQUELAE

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This case is presented to the staff for confirmation of my findings, and with the view of eliciting a discussion as to the prognosis with especial reference to the persistence of a symptom-complex that I have been unable to find described in the literature, in so far as such literature was available to me.

Whether or not such a sequence in epidemic encephalitis has ever been described at all, I do not know; if it has, its rarity justifies my obtruding another article on the already overgrown volume of writings on this affection.

When Economo of Vienna coined the peculiar term "encephalitis lethargica," he did not really describe a new disease picture, but his appellation persisted principally on account of the war, and the lack of communication between various medical channels and centers. The British had had an epidemic of some unusual conditions that they had described under various captions, such as epidemic botulism, epidemic drowsiness, epidemic intoxication with lethargy; and it seems that Marinesco of Roumania came to the London hospitals to study the epidemiology of these affections. At this time the British themselves began calling their cases "encephalitis lethargica." This seems strange, not only that there was such animosity in England against the enemy, but since the days of Hughlings Jackson the British were never prone to accept any too quickly terms with Teutonic terminology.

The disease under different names was known to the older writers and among more modern writers one finds a description, with a bibliography by Oppenheim, in Nothnagel's Handbuch. Here in New York I find two very interesting cases described twenty years ago in the *Medical Record,* and *Medical News,* one by Dana and Schlapp, and the other by Harlow Brooks and Theodore Janeway. In the light of our present day knowledge one can see that these cases were probably cases of encephalitis lethargica although they were not called that in those days. Both were carefully autopsied and showed what we would find and describe to-day. In Brooks' case a nineteen year old girl was taken to his service at St. Vincent's Hospital unconscious, groaning, with paraplegia, double Babinski, drop foot, and remained in a drowsy state for two weeks and died. It was clear that she had some noninflammatory disease of the cerebral cortex causing lethargy. The autopsy by Janeway showed no gross lesions, and blood cultures were negative. Microscopic sections revealed that the lymph spaces of the pia arachnoid were greatly dilated, and contained serous exudate with many lymphocytes. Connective tissue cells were numerous, and their nuclei showed much evidence of proliferation. Blood vessels were moderately congested, their walls not thickened, they looked normal. Cortical vessels were normal, there were slight congestion and perivascular lymph spaces almost universally dilated. That is about all that one finds in our cases, with the exception that Strauss and Loewe have recently isolated an organism.

After Economo drew attention to this condition, reports began to come in from all parts of the world, from Australia, from Netter in Paris, Bassoe, Chicago, and here in New York clinical studies from Mt. Sinai Hospital by Abrahamson and Strauss, and from the Presbyterian Hospital by Tilney, Riley and Howe. From all these sources a wide range of symptoms can be gathered, as wide as the anatomical possibilities, in the brain, medulla, spinal cord, and even endocrine involvement, but primarily in the midbrain.

It seems to me that besides the classification of the enormous number of clinical symptoms, from an anatomical basis, there can be made another division according to the time and nature of the infection. The symptoms thus group themselves into four grades or epochs: (1) Those that occur mainly, if not entirely in the stage of incipiency; (2) those of active involvement; (3) the stage of disease with decline (if there has not occurred an exitus lethalis), and (4) the stage of sequelae. So that such signs as lassitude, diffuse pains, even local pain in the head or abdomen with gastroenteric symptoms, respiratory infection, nasal, and lachrymal infections, may constitute the stage of incipiency, wherein the patient may or may not be drowsy. In fact many cases, such as the one I am going to present, suffer from insomnia. In this stage the diagnosis often cannot be made. It can be suspected however, especially in view of an epidemic. The second stage, however, is a reality, the patient is no longer ambulatory, and his apathy or mental indifference is already attracting attention of his attendants. Gradually cranial nerve weakness if not paralyses makes its appearance. There is now no doubt as to the diagnosis. In the third stage there is a gradual recession of all the main symptoms, some in part, some altogether, so that diplopia, usually very distressing in the incipient stage, now generally disappears. If the patient dies, it is usually at this period of the disease; if not, he passes into a fourth stage, wherein he is out of danger of death from exhaustion or complications, is apparently well, and now beginning to show new signs, in the nature of sequelae.

Signs that might have been very mildly apparent, or usually not present at all, now make their appearance, and the peculiar part of it is that these symptoms that are last to appear are most apt to stay longest, possibly permanently. I refer to such movements as paralysis agitans, or athetoid-like contractions that appear at this time. The reason I have dwelt upon this in a rather lengthy preliminary is that my case shows signs of vagus nerve affection, and that the change in rate and character of the respiration came on in this fourth stage of the disease, and if not permanent (which it threatens to be) will last for a goodly length of time.

This boy is eight years old, the second of two children, the other being well. The mother had given birth to four full term children that looked normal, but that could not be gotten to life. The story was that the doctors could not start respiration. I saw the last one myself five years ago, the infant lived about eighteen hours, had persistent cyanosis, unless slapped, or immersed in cold and warm water, alternatingly. I did a postmortem and found nothing abnormal anywhere until I reached the kidneys. These were tremendously enlarged, nearly the size of my fists, and sections made by Dr. Cornwall showed that they were the seat of congenitally acquired large cysts. I mention this because it gave the first clue to the cause of the stillbirths and the respiratory difficulty, and also because the obstetrical history of the mother points so strongly to syphilis. That there is no syphilis I know, for several Wassermanns, on the fetus, mother and father of this boy were negative. The boy has no clinical signs that are suspicious of syphilis. This lad was always well, developed normally, has good habits, and is very bright, in and out of school. About nine weeks ago, during the influenza epidemic, he had some mild headache, and general malaise, and it is said he had an elevation of temperature. He remained at home from school but one day, although he says he did not feel well, even when he returned to school. About one week later, he was out sleighriding one afternoon, and went down hill on a sled lying on his abdomen. Suddenly, as he approached the center of the road, an automobile made its appearance, and the boy was picked up from under its front wheel. He was not hurt, but dreadfully frightened. He came home, trembled and cried constantly for hours, and was too scared to take food. He went to bed, but did not sleep, and in the morning refused to go to school, a very unusual situation for this lad. There seemed to have been effected a change in his mentality, he was stupid, and responded to commands very superficially. Although he showed no signs that were suspicious of his having delusions or hallucinations, he was evidently not normal. He would go to the sink, turn on the faucet, with the purpose of getting some water, he would stand there, or turn around and go slowly to bed, forgetting to turn off the faucet. He would stare into space, and it was with difficulty that his family could get him to respond to questions. On being coaxed or threatened he would say, "I am too tired, please don't bother me, that is the best that I can do, I feel exhausted."

The family, and the family physician took the condition to be one of hysteria, and traced the cause of the boy's symptoms to his fright, when he ran into the automobile. After several days, I was called and found the boy in bed, spine completely flexed, hands on his head, whining. There was apparently very severe pain to palpation in the ligamentum nuchae region. This was general, but more severe on the right side. There was no retraction of the head. It was at once apparent to me that the history as given was misleading, and next to worthless. The only clue that I got came from the boy himself, who told me that he had had diplopia on six or seven occasions, after that day of so-called influenza. He said the diplopia began in the morning, during school hours, and that it usually wore itself off at about noon or early afternoon. I began to look for early signs of encephalitis lethargica, but there were not any. But he showed from the next day on, for about a week, constant and progressive cranial nerve involvement. The first was a mild left facial. This was interesting in that it was a "thalamic facial" as Dr. Casamajor, who saw him with me, called it. The middle distribution was the most affected. The patient showed no difference between both sides of his face when he was told to smile. But when he smiled without being fully conscious of the act, his left face stood out in true palsy. He had involvement of his third, fourth, sixth, ninth and eleventh nerves to a mild degree, all on the left side.

He passed into what I have called the third stage of his disease, in the second week of his illness, and his symptoms all cleared up. His diplopia and torticollis gradually left him. He had had a mild elevation of temperature, to 101°; this came down to 99½°, to 100° by rectum, and hovered near the normal, but very slightly above, for many weeks. His respirations were about 20 per minute, his pulse about 90. He was apparently well, mentality normal. He showed, however, a very mild form of ataxia in his left upper and lower extremities. It was just a little more than a suspicion of ataxia. It was now that he passed into what I have called the fourth stage of the disease. Here was a boy all well developing something new, possibly permanent symptoms, at a stage of the disease when it was safe to say that he was over his illness. His cranial nerve conditions cleared up, drowsiness left, pulse, temperature, respiration became normal, appetite was good and it was with diffidence that his family kept him in bed. Then his ataxia gradually began to increase, and with it came rigidity of the neck, and a rather fixed head, so that he looked typically Parkinsonian. It was noticed that his respirations were rather rapid, and I was sent for. I found him breathing very deeply and rapidly, the rate was about 45 per minute. At first thought I looked for pneumonia. His chest was free of any râles or pathological signs, his temperature was normal, or elevated only half a degree. His pulse was 100-110. Examination of his blood and urine was entirely negative. For seven weeks his respiration has been rapid. You can see as he sits here now how rapid it is; it is to-day a little better, being about 38 per minute. But I have seen him at all hours during the past seven weeks and it is usually 45 per minute. During the past two weeks it has become somewhat irregular, and there is a catch to his breathing somewhat like Cheyne-Stokes. His breathing is decidedly worse during sleep; it is a little faster, and more jerky. Graphically represented I would say that his respirations are dichrotic, and with two wavy jerks in inspiration. His larynx and pharynx are normal to laryngoscopic examination, yet they are the seat of roapy mucous accumulations in the morning. The depth of his breathing and the rate are slightly amenable to re-education, and I think that his nurse may succeed in improving his respiration to some extent if she persists in her efforts at breathing exercises with the lad.

From repeated negative examinations of the chest, blood, urine, and the absence of temperature, one has a right to suspect that the disturbance of respiration, going on for seven weeks, is central, or neuritic in origin, and that it is probably nuclear. Whether unilateral, or bilateral, I am unable to say. What will become of his rapid breathing? Will it very gradually decrease in rate, or is life compatible with this new system? What will be the effect on his metabolism? Personally I confess that I have never heard of a persistent tachyrespiria. And are we to look forward to any changes in the way of compensations? One thing seems certain, that but for the fact that he has a little hyperhidrosis, it doesn't seem to cause him any fatigue. If anything he has gained in weight, and strength.

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DELAYED DIAGNOSIS IN A CASE OF EPIDEMIC ENCEPHALITIS

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The patient, T. S., History No. 29931, twenty-eight years of age, Austrian by birth, residing in the United States sixteen years, is at present engaged as a terra cotta worker. He came to the Vanderbilt Clinic, Neurological Department, October 15, 1920, complaining of nervousness, weakness, insomnia, stiffness and tremor. These symptoms have been marked for the past four months. The onset of his illness has been very gradual and has extended over a period of approximately seven years.

Sixteen years ago the patient came to the United States and secured employment in Pennsylvania as a miner, at which occupation he worked for six years. He then did a laborer's work on a railroad for six months. Following this he became a terra cotta worker, continuing at this for six and a half years. At some time during this period he noticed that his right hand trembled when he signed his name, and that his writing had changed. This trouble, however, did not interfere with his work. Between 1916 and 1918, for a year and a half, he worked in a copper factory where shells were manufactured. From March, 1919, until two months ago he worked at terra cotta again.

Seven months ago, he began to suffer with severe headaches and burning sensations in his head, varying in position from the top of the head to the sides, back of the head and neck. The severity of the pain was such as to prevent sleep during the greater part of the night. A tremor then delevoped in the right hand and gradually involved the right leg. According to the patient this tremor is present while he is at rest as well as in motion. A general rigidity and weakness was next noticed. The stiffness was limited not alone to the extremities but also involved the face. With the increase of rigidity and tremor, the headaches ceased and the insomnia gradually disappeared. His weakness is such that he easily becomes fatigued on walking and prefers to lie down. He has also noted that his speech is slower and changed and that the tone of his voice is lower. In one and a half years his weight has increased from 143 to 168 pounds.

Past History.—Measles, pneumonia, malaria (?). Venereal diseases denied.

Habits.—Negative.

Family History.—Mother died at forty-six years from cardiac disease. Father living and well. No similar illness in family.

Marital History.—One child three years old, living and well. No miscarriages.

PHYSICAL EXAMINATION

VOLUNTARY MOTOR SYSTEM.—*Abnormal Attitudes and Deformities.*—Masklike features, sits very rigid, hands at sides.

Gait.—On toes, on heels, on a line, backwards, sidewise, running, all normal. Gait slow but steady with arms slightly flexed.

Associated movement of trunk, normal. Associated movement of right arm and hand gone. Associated movement of head, normal.

Coordination.—Right-handed individual. Equilibratory: Standing with feet together, eyes open and closed, normal. Non-equilibratory: Finger-to-nose, eyes open and closed, normal except for tremor. Finger-to-finger, normal except for tremor. Toes-to-object-above, recumbent position, eyes open and closed, left and right, normal except for tremor. Heel-to-knee and along shin, eyes open and closed, left and right, normal except for tremor.

Adiadochokinesis absent. Handwriting displays tremor. Speech, slow, tone lower. Test phrase, "Third riding artillary brigade," fair.

ABNORMAL INVOLUNTARY MOVEMENTS.—Coarse, medium tremor of hands, right greater than left, intensified by motion, absent when at rest. Tremor in lower extremities, right greater than left. All movements slow. Coarse rigid tremor of lips when tongue is protruded. Fine rapid tremor of tongue.

Reflexes:

DEEP

	Left	Right
Biceps	2	2
Triceps	1	1
Patellar	2	2
Achilles	2	2
Ankle clonus	0	0

Right reflexes apparently not so easily elicited.

SUPERFICIAL

	Right	Left
Supraumbilical	2	2
Suprapubic	2	2
Upper lateral abdominal	2	2
Lower lateral abdominal	2	2
Cremasteric	2	2
Babinski	0	0

Muscle Strength.—All motions of flexion, extension, abduction, adduction, supination and pronation, opposed and unopposed, are normal except that on passive movement of abduction of lower extremities, more on coarse tremor is present.

Muscle Status.—Normal except for general slight rigidity of extremities, more on right side, least on left upper. Slight cogwheel release on right at times, especially on lower right.

Abnormal Associated Movements.—None present.

GENERAL SENSORY.—Touch, acuity, localization, discrimination, normal.

Pain, superficial, apparently felt rather more keenly on the right than on left lower extremity.

Muscle tendon of extremities, normal.

Stereognosis, normal. Barognosis, normal.

CRANIAL NERVES.—Optic: Fundi normal, pupils equal, medium size, round, centrally placed, react to light and accommodation, no hippus, no nystagmus.

The 3rd, 4th, 5th and 6th cranial nerves, normal; 7th cranial nerve, very difficult and usually impossible voluntarily to wrinkle forehead. Tremor of lips when showing tongue. Limited smile and laugh.

Associated movements of face, eyes and head, limited; 8th, 9th, 10th, 11th cranial nerves, normal; 12th cranial nerve, normal except for fine tremor of tongue. Tongue coated. Movements of tongue normal.

MENTAL STATUS.—Intelligence, emotions, attention, memory, normal.

SYSTEMIC.—Except for enlarged tonsils and palpable anterior cervical, axillary and inguinal nodes, and pyorrhea, the physical examination is negative. Pulse 72, regular, rhythmical and of good quality. Blood pressure: systolic, 130 mm. Hg., diastolic, 80 mm. Hg. Temperature 98°.

LABORATORY FINDINGS.—Wassermann: negative. Urine: negative including examination for lead. Blood smear for basophilic stipplings: negative.

COMMENT

This case is interesting because of the delay in the final diagnosis and also because the type complex so overshadowed the original disease. From the point of view of prognosis the differential diagnosis is, needless to say, very vital.

When the patient presented himself at the Vanderbilt Clinic, some of us felt, tentatively, that the case was obviously one of paralysis agitans. However, before arriving at our final diagnosis, the other possibilities were considered, and as not infrequently happens, one of the remote possibilities became the true diagnosis.

The possibilities considered in this case were, first, paralysis agitans, second, metallic encephalitis, third, epidemic encephalitis of the paralysis agitans type.

In considering paralysis agitans, we must place the inception of the disease seven years ago, when the patient first noted a change in his writing. Assuming this to be true, then we should expect many of the typical signs of the disease to be present, namely propulsion, retropulsion, pill-rolling and rest tremor. The masklike appearance, rigidity, weakness, cogwheel jerk as felt in the right elbow and the loss of associated body movements would indicate a paralysis agitans type of disease and not a true Parkinson's.

The possibility of a metallic encephalitis is rather remote as but little lead is used in the glazing of the clay of terra cotta. Lead encephalitis is accompanied by arteriosclerosis, and as pointed out by Casamajor the encephalitis is only secondary to the arteriosclerosis. Lead causes the arteriosclerosis and the latter the cerebral degeneration. As no evidence of arteriosclerosis was present, we can dismiss this consideration.

An illness, acute in onset, with severe constant headache and insomnia occurring during the encephalitis epidemic and having residual paralysis agitans symptoms, speaks strongly for an infectious disease. It is not inconceivable that the patient passed through a very mild form of epidemic encephalitis of the paralysis agitans type which did not confine him to bed.

The fact that the headaches and insomnia improved as the rigidity and tremor became accentuated may be interpreted on the theory that it was not because of the disappearance of the headaches and insomnia that the rigidity and tremor became marked, but rather because of the accentuation of the tremor and rigidity that the headaches and insomnia disappeared from the attention of the patient.

In carefully studying the evidence at hand we are inclined to believe that epidemic encephalitis is the correct diagnosis. Time, and improvement of the condition, will either prove or disprove our contention.

A CASE OF HEMI-DYSTONIA*

Dystonia Musculorum Deformans

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The subject of dystonia musculorum deformans has been so thoroughly covered since Ziehen, Oppenheim, Flautau and Sterling and subsequent authors in classical descriptions enriched organic neurology with this distinct clinical entity, that extended introduction of this phase of the subject seems superfluous. The original observation of Ziehen, who regarded the affection as a neurosis, was disproved by later writers who considered the disease as a fundamental, organic affection of the central nervous system. This has been amply proved by Thomalla who in autopsy material from one of his cases found degenerative changes in the globus pallidus portion of the lenticular nucleus. The presentation of this case as a hemi-dystonic type of affection is purely arbitrary, as eventually a bilateral involvement will occur evidence of which is already manifest. The contrast between this case and the one presented before the conference several weeks ago (in which the entire bodily musculature was convulsed in a relentless dystonia) is so striking that for the present at least it seems permissible to characterize it as a hemi-dystonia in view of its sharp limitations to one half of the body. It is quite obvious that there are defects in such characterization both from the viewpoint of an overcrowded neurologic nomenclature and the difficulty in maintaining one's thesis that the affection as viewed clinically is strictly unilateral.

The Case.—The boy is a native of Russia, of Hebrew parentage and is fourteen years of age. He came to this country seven years ago and was perfectly well up to within four years. Then he noticed that his left leg commenced to turn in and this was followed, several months later, by a twisting and turning of the head and left upper extremity. Before the head and arm became involved however, he visited a clinic where a cast was applied to his foot. This was removed at the end of three months and a flexion contracture of the large toe was found. Operative procedures were promptly undertaken to relieve this condition, which in view of subsequent developments were manifestly a failure. His previous history, prior to the development of his affection, is without incident. There is no history of any acute illness or trauma. He attended school regularly for three years and then discontinued on account of his difficulty in getting about. His work at school was satisfactory to his teachers.

Family History.—The boy's parents are dead, one dying of pneumonia and the other of acute infection of the leg. A sister, aged seventeen, is living and presents no neurological abnormalities.

Voluntary Motor System. *Abnormal Attitudes and Deformities.*—The patient's head is drawn toward the left shoulder, the contraction being so severe at times that quite frequently the head rests on the shoulder joint. The left hand at the wrist is held in a state of flexion contracture in which the fingers partially share. At times the forearm is held completely extended, then, again, there is a marked flexion at the elbow suggestive of a hemiplegic deformity, the alternate contractions and decontractions determining the variability in the normal postures. In the lower extremity there is a marked flexor spasm of all the toes and the pes cavus suggests the ultimate "semi-lunar" foot.

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* Presented at a clinical conference June 17, 1920, from the Hospital for Deformities and Joint Diseases. Service of Dr. H. W. Frauenthal.

Gait.—This is quite bizarre. The head is held in extreme flexion to the left, the left upper extremity in tonic extension. With each forward step there is simultaneous, forcible extension of the left leg and a violent lateral flexion of the trunk. The movements are decidedly stereotyped and do not materially interfere with the phases of progression. The pelvis, in a rather mild degree, shares in the dystonic phenomena.

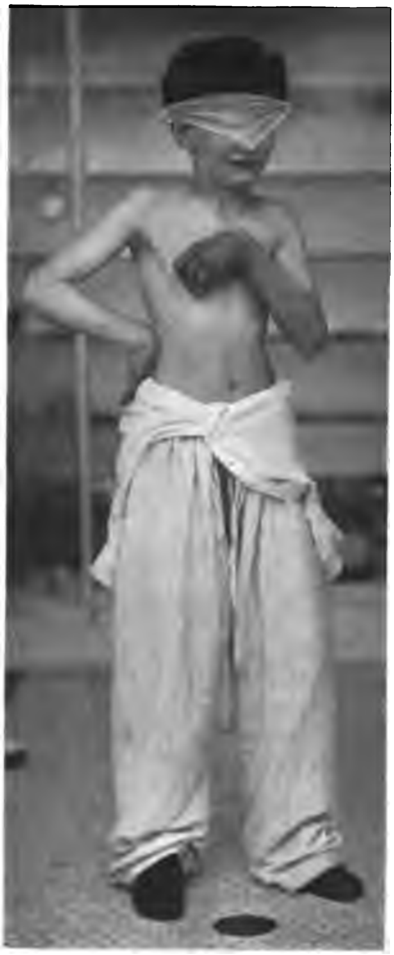

FIG. 1. HEMIPLEGIC ATTITUDE IN HEMI-DYSTONIA.

Apraxia.—The performance of purposeful movements is somewhat interfered with in view of the disturbance occasioned by the muscle unrest. They are performed, however, in a tolerably satisfactory manner without assistance.

Abnormal Involuntary Movements.—When the patient is lying or sitting down there are moderate movements of the head and choreo-athetotic movements of the left upper extremity. This abnormal muscle activity reaches its maximum when the patient is standing up or walking. Under these conditions the muscles become violently active, going through all the phases of tonus disturbance and assuming characteristic clownish bizarre contortions and attitudes. This curious muscle behavior may rightly be called a hemi-dystonia in view of its sharp limitations to the entire left side.

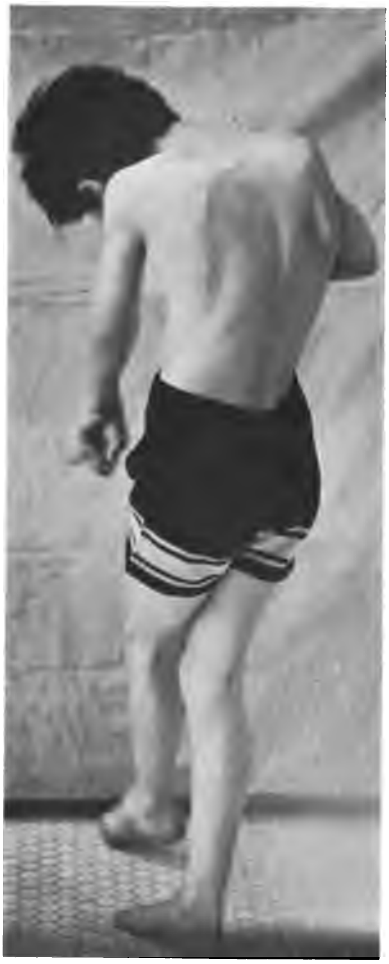

FIG. 2. DYSTONIC ATTITUDE IN "HEMI-DYSTONIA" MUSCULORUM DEFORMANS.

Coordination.—All coordinative acts are performed with very little difficulty. At times their smooth performance is somewhat marred by the involuntary muscle activity, but in the main there are no gross disturbances noted.

Reflexes.—The superficial reflexes are easily elicited and the responses are prompt and equal. The deep reflexes, however, present a problem which in view of the hypertonicity cannot be overcome. Numerous efforts made to elicit them on the affected upper and lower extremities are without success. One gains the impression, however, that if the hypertonus could be temporarily overcome reflex activity would readily manifest itself. The deep reflexes in the right upper extremity are elicited after considerable patience and effort. The right lower extremity readily responds to tendon stimuli. Pathological reflexes are not obtained.

Muscle Strength.—There is apparently no impairment noted in all types of muscular effort. All types of voluntary muscle activity are performed in a prompt and vigorous manner.

Abnormal Associated Movements.—There are no demonstrable, abnormal associated movements.

Muscle Status.—There are no hypertrophies or atrophies present. Hypertonicity is present in the left upper and lower extremity. The right upper extremity is moderately resistant to passive movements but this hypertonus is not as marked as in the above mentioned extremities. The electrical reactions show no variation from the normal.

NERVE STATUS.—There are no demonstrable abnormalities noted in the peripheral neural apparatus.

SENSORY EXAMINATION.—This shows perfect acuity in localization and discrimination in all types of sensory reactions.

CRANIAL NERVES.—The cranial nerves were carefully investigated and found functionally intact.

LABORATORY EXAMINATION.—All laboratory tests have yielded uniformly negative results.

MENTAL STATUS.—The patient is bright, intelligent, cooperates readily in the examination and apparently shows no emotional or psychotic disturbances. He displays no anxiety or distress and views his condition with considerable equanimity. Mention is particularly made of this mental placidity, as it seems to be a marked trait in the cases observed in spite of the ceaseless motor agitation. The mental attitude appears to be one of resigned tolerance toward an inescapable condition. Obviously the age of the patients and their freedom from the perplexities of life must determine, in a large measure, their attitude toward the affection.

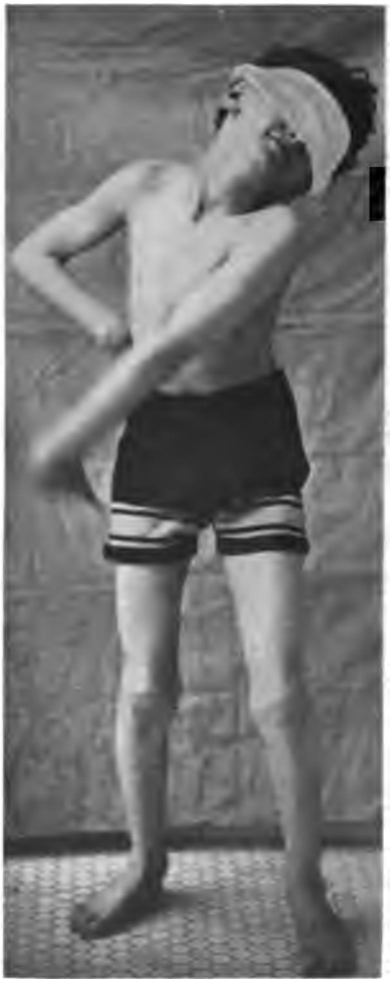

FIG. 3. DYSTONIC PHENOMENA LIMITED TO HEAD AND LEFT UPPER AND LOWER EXTREMITIES (HEMI-DYSTONIA).

CONCLUSIONS

An analysis of the syndrome presented by this patient reveals a history of onset at an early age, with the characteristic abnormal muscle behavior limited to the lower extremity. In a study of the cases reported in the literature and those personally observed, this was the invariable mode of onset. In no case was the upper extremity first affected. Curiously enough most of the cases were subjected to orthopedic measures in the beginning, and the case here presented did not escape the application of a cast and subsequent operative procedures. Obviously these measures proved to be valueless. The invasion of the trunk, arm and head followed soon after and apparently has been limited to these areas for the past three years. The invasion of the other side seems imminent in view of the perceptible athetotic movements present in the right upper extremity. Many years may elapse, however, before an invasion of the opposite side occurs as pointed out by Hunt. The uniform mode of onset and the limitation of the abnormality to one side for an indefinite period may serve as a ready method of differentiation from other affections that have choreo-athetotic manifestations. To enter into a freer discussion of the various affections that are characterized by these bizarre and curious movements is not the purpose of these notes. Suffice to say that Thomalla has attempted to group all degenerative affections with choreiform and athetotic movements into the more comprehensive designations of dystonia lenticularis. Inasmuch as the localization and pathology of affection with dystonic phenomena, according to recent investigators, seem to differ but little, one cannot but feel that this designation bears considerable merit. At any rate it will tend to clarify the confusion created by the various types of abnormal involuntary muscle movements and give them a common localization and pathology.

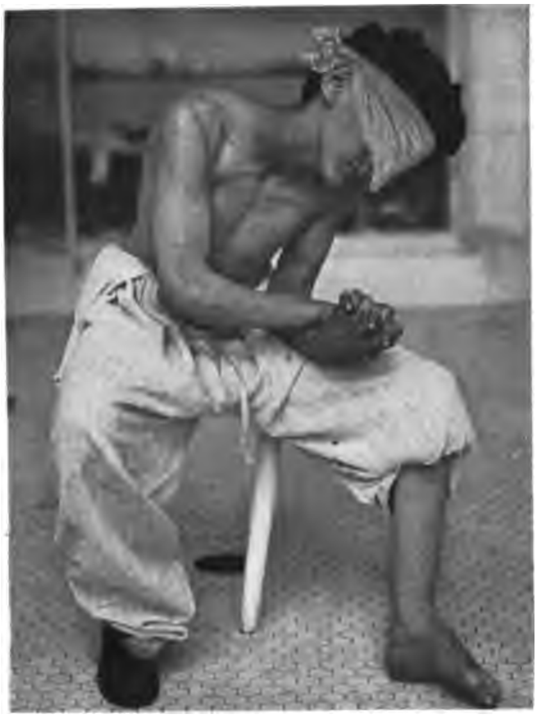

FIG. 4. EARLY APPEARANCE OF ULTIMATE "SEMI-LUNAR FOOT.

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THE SOCIAL DEVELOPMENT OF MAN AND HIS PROBLEMS OF ADJUSTMENT

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While it might be doubted that psychiatric difficulties are increasing in the present day, yet it is conceded that they have increased in number in the past two hundred years. As far back as history records there have always been neurotics. Probably they existed, in small numbers at least, before the dawn of history, for in all races civilization was fairly well advanced before the development of written speech. The cause of the neuroses has been a matter of interest to medical writers since Hippocrates, but only within the last quarter of a century has the importance of the psychogenic factor been universally recognized.

The neurosis to-day is looked upon as one method of solution of the personal problems which beset the individual living in a civilized community. He becomes neurotic because he is at the same time an individualistic and a socialized animal. The development of such a condition has been a slow and gradual one and a part of the general development of the human species. It must not be understood by this that man has developed into a contrasting, individualistic and socialized being as the result of his evolution. He has always been such; but the social and physical features of his problems have been so pressing that the individual problem could not become the more prominent. Only when these other problems had been solved in a manner more or less satisfactory did the individual problem appear as most urgent for solution. The individual problem has always existed, and many of the social problem solutions have been reached in an attempt to attain the pre-eminence of the self.

The principal problem of life is living, and to this may be added a second biological necessity—propagation, which is a habit of protoplasm. The earliest multicellular forms succeed in this struggle with their environment by two means: profligate propagation in the hope that a few may survive, and the development of protective structures. The former method may have been satisfactory to the species, but only the latter could satisfy the individual. Protective structures arose from the ectoderm, first in the form of shells and later in the development of the nervous system. The latter has proven itself by far the most efficient method. The development of special sense organs and the distance receptors for smell, hearing and sight gave the animal a better knowledge of his environment and allowed him to learn how to cope with it. Incidentally, it made life more worth living. Many forms persisted in struggling with this better-known environment with the aid of ectodermal defense structures—shells, hide, hoofs, teeth, claws, and horns. In others, the mesozoic reptiles, this end was attempted by the acquiring of enormous size. This method of nature was tried and found wanting. Man, says Langdon Brown, has attained his position at the head of the phylogenetic series simply because he has never allowed himself to be led away from the development of the nervous system.

Here we see the first problem of life—the struggle with the physical environment. More means than I have indicated have been attempted by species to solve this problem; and while many forms found that life became possible by such means, yet the means themselves necessarily restricted the individual and made further progress extremely difficult. Long before the advent of man, even before the vertebrates, a solution had been found by certain insects in the swarm, which has its counterpart in the vertebrates—the herd. While the herd solves, more or less efficiently, the problem of physical environment, it does it at the expense of the individualistic tendencies. In the swarming insects, the ant and the bee, the individual is entirely submerged in the welfare of the herd, even to the extent of completely losing sex. Such complete denial of individualism is not demanded of the vertebrate herd animals, yet the herd restricts the individual for its own welfare. In the herd, according to Trotter, selection by survival of the fittest is transferred from the individual to the group. The individual survives not because of his own fitness, but due to the fitness of the group to which he belongs. While this is of distinct advantage to the individual, still it imposes herd rules on him which are inimical to individual development.

When man appeared on earth the herd idea was already well developed for him, for it had been the habit of his ancestors for centuries. Thus the problem of the struggle with the physical environment had been solved. With his more highly developed nervous system and superior intelligence, the urge of the individualistic tendencies became greater, and with further development and the increase in the size of the herd he came face to face with the second great problem of his evolution—the struggle with the biochemical environment, i.e., nutrition. When the herd animal is dependent on the hunt and on browsing for its food, the size of the herd is restricted by the amount of food available. The available food may be increased by increasing the strength and skill in hunting animals and by increasing the speed in herbivorous animals, thereby increasing their feeding range. But this ultimately has its limitations, and in any event it forces the animal to spend too much of his time and energy in the simple pursuit of food to allow for individual development. Man must have found some better means than these to solve this difficulty or he could not have progressed further. The means he found were husbandry and agriculture, the care of domesticated animals and the growing of vegetable food. By such means the limits of the size of the herd were raised without interfering with the individual progress of the members. Some groups remained nomads with their strong fleet-footed cattle, while others, finding fertile valleys where crops were easily produced, settled down to an easier farming life which allowed for other activities besides simple protection from death from enemies or starvation. This was notably true in Mesopotamia, the cradle of civilization.

When man was no longer obliged to spend all his time in pursuit of food he began to utilize his spare time in individual development. His societies—for such we can now call the herd—increased rapidly in number. These numbers increased the security of the group from outside enemies and made agriculture easier. Probably the first attempt at individualism was in the formation of the family and the idea of private property. The first private property consisted of stone tools and instruments. These favorite bits of stone belonged to the individual and frequently were buried with him. They are to be found in primitive graves to-day. The family was part of the property, first of the mother and later of the father. Such a system allowed of considerable individual development, and as the development progressed inequalities of station arose in the group. Such inequalities hindered the individual development of the younger, the less favored and the less fortunate. Thus man found himself with a new problem to meet: the problem of the struggle with his fellow man; the struggle for equality of social position and advantages; the struggle for the right to develop individually which continues to-day.

Time does not permit me to enter into the struggle in the family, between father and sons, which has played such an important part in the development of the human personality and character. One can but outline the main attempts at solution of this struggle. It is the record of history, the attempt of individuals and groups to develop individual superiority and resulting ease of living at the expense of others; the attempts of these others to develop their own individualities by the overthrow of their oppressors. Physical might marked the first of these attempts. Strong tribes conquered the more civilized weak tribes and forced them into slavery. Thereafter the strong ruled in ease, with the slaves doing the hard work, and the conquerors absorbed the civilization of the conquered only in their turn to be conquered by still stronger races. During this time communities were increasing in number and cities arose where life was safer and more luxurious and possibilities for individual development were greater. More and more individual interests clashed among the rulers and among the slaves and something besides physical might was necessary to solve this problem of man's struggle with his fellows and allow individual development further to progress. Hence, rules for the game of living arose, rules which we call laws. With the advent of laws, an enforcing agency became necessary and this agency was government. It seems that the human mind cannot start anything new without first going backward to something it knew before—a precedent. So the earliest form of government was modeled on the early family under control of the father or strongest man, who became the chief or king. The king in history has had a varied career, sometimes ascending in favor, sometimes being replaced by autocracies or democracies. Even to-day the human mind clings closely to the idea of the father-king. Rome passed from a democracy to an autocracy and then to an empire under the Caesars. As Wells points out, this "Caesar idea" has persisted and the war of 1914–1918 deposed four Caesars, the Kaisers of Germany and Austria, the Czars of Russia and Bulgaria.

During all this time the social struggle, the struggle of man with his fellows, continued. Governments as evolved did not completely solve it; nor is it solved to-day. There is not enough opportunity in the world to satisfy the selfish. It is still believed that one man can rise only at the expense of others. Those at the top must find some means to meet the individualistic and class pressure from below. Cruelties sufficed for a time—that old resort to physical might. These gave place, eventually, to compensations for the lower classes and oppressed individuals. The greatest of these compensations was religion.

Religion, and especially Christianity, has ever been a solace for the oppressed. Christianity in Rome was considered a "good religion for slaves." Loyalty to the God-King was another solace, a virtue which was its own reward; and out of this has grown patriotism.

England was responsible for the next great advance in government, representative government. At the end of the eighteenth century, popular government came to the fore principally through the revolutions in America and France. By this means special privilege was to be wiped out and each member of the state given the right to develop his individuality equally. The preamble to the Declaration of Independence contains this old fallacy: "We hold these truths to be self-evident, that all men are created equal, that they are endowed by their Creator with certain inalienable Rights, that among these are Life, Liberty and the pursuit of Happiness." This was not a new idea. It is only what man has been struggling for since the first formation of societies. To attain this goal man continues to make all kinds of experiments in government. No scheme is too absurd to be tried provided only that this aim is to be kept in view. The present experiment in Russia is but one of the more radical of these experiments.

Whether or not man will ever attain this end, it is nevertheless fairly evident that he has come very close to it in the modern civilized state under the republican form of government. He is still experimenting with laws in the attempt to solve every ill of society, till we are cursed with too many laws. Laws change many things but do not change the nature of man. Under present conditions he is comparatively safe and comfortable and is given ample opportunities to develop his individuality. Now that man is free to take up the problem of his individualistic development, he finds himself face to face with a new problem the importance of which he is just beginning to realize. This is the last of the great problems of man—the problem of man's struggle with himself.

We must set out with the proposition that man started with a nervous system and especially a cerebral cortex probably very much like what we have to-day. Stable civilization probably began about 10,000 years ago. That is probably the date when man first approached the problem of his struggle with his fellow man, a problem for which he has only recently found a fairly satisfactory solution. Counting three generations to a century we find but a little over four hundred generations have elapsed since the beginning of civilization. One may well wonder whether such a number is sufficient to alter materially the nature of man. Intelligence has enormously increased, due to the printing press and literature, and it is likely that what change has occurred in man's mentality has been almost exclusively in the intellectual field.

Man's emotions are still primitive and his intellect is still in great measure under the emotional sway. Certain inhibitions have been learned, or rather have been forced on man by his social environment; but this so-called control is a very superficial thing. It is this development and elaboration of the unconscious that has made social progress possible. One might say that it was the discovery of the possibility of using the unconscious as a repository for purely selfish individualistic desires that made man a civilized social animal. The denial of the existence of the unconscious made further advancement possible.

It is this denial of the unconscious that has forced on man the struggle with himself. The glorification of the conscious as the only part of the mind resulted in man's taking a more elevated view of himself, and with the repressions and resulting sublimations, social progress went on by great leaps and bounds. Man tried to make this, his world, a better place to live in, with great success. The enormous social betterment of the past century and still more the mastery over nature and the elements, of the arts and science, has made for us a much more comfortable place.

But the human mind has not kept pace with this social and political advancement. The intellect has overstepped itself. The rest of the mentality has not caught up. Hence, we note a constant tendency to regress. When struggles become great, when new and difficult situations are to be met, the tendency to slip back is seen, the return to a lower level for solution, the search for a precedent. Such regressions are seen in large and small groups and in individuals. The late war was a regression for almost the entire world. Hardly less universal was the post-war revival of spiritualism and the Ouija craze. The New York papers of December 20, 1920, contained the story of an old woman in Pennsylvania who appealed to the police for protection against her neighbors who claimed she was a witch. They had accused her of being the cause of the death of their pigs, cows and horses. The Salem witch craze in 1645–92 was clearly a regression, and Potts speaks of more modern witch crazes. The mind in Pennsylvania in 1920 can be very little different from that in Salem of 250 years ago.

Regressions in individuals are still more common. This is the psychoneurosis which is increasing in our society. Freud has shown how accurately the symptomatology of many neuroses corresponds to the beliefs and fears of primitive man. Among the psychoses these regressions both to the infancy of the individual and the infancy of the race have given us a large literature. That personal difficulties can be settled by mental slip-backs to primitive modes of thinking is what gives us our problem to-day.

So to-day we are entering into a third stage of man's development—the stage of man's struggle with himself. An age long, he has been engaged in his struggle with his fellow man, and now the solution is within reach. He has reached this solution by a remarkable intellectual development in the conscious field, leaving his emotions and the unconscious in a state of pristine savagery. These parts of the mentality have now to grow up to the other before further progress and happiness are possible. The question is: Can man do this? Now that he has gotten the plaything he has cried for for ages—"the right to Life, Liberty and the pursuit of Happiness"—has he the mentality to play with it?

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THE SUBSTANTIA NIGRA: A BRIEF COMPARATIVE ANATOMICAL STUDY*

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INTRODUCTION

Although there is comparatively little positive knowledge about the substantia nigra quite a great deal of literature has been accumulated on the subject and not a little research has been undertaken both as to its structure and function. Vicq d' Azyr was the first man to describe the substantia nigra under the name of "locus niger," then came Soemmering, whose name it bears, and finally Meynert called attention to its ganglionic character. Because of its striking appearance even to the naked eye and because of its pigmented nature a number of observers have studied the substantia nigra with a view of determining its fiber connections, its cell structure and its physiological function. Most of the work on the cells has been done by the Italian school, particularly by Mingazzini, Mirto and Amaldi. A great deal of the work on the fiber connections was carried out by German investigators, more especially Meynert, Economo, Obersteiner, Monakow, Kölliker, Werdnig and a number of others. The physiology has been investigated by the Russian school in Bechterew's laboratory by Jürman, Witkowski and Muratow. More recently Julius Bauer has undertaken quite an extensive comparative anatomical study of the substantia nigra in man and the primates, the prosimians, chiroptera, carnivora, pinnipedia, insectivora, rodentia, proboscidia, ungulata, natantia, edentata and marsupialia. Bauer also studied the pigment in the cells of the substantia nigra and extensively reviewed the literature. (Many of the quotations in this article are based on Bauer's work, as also on those of Bechterew, Obersteiner, Monakow, etc.)

At first unaware that so much excellent work had been done on the substantia nigra the writer was tempted (really through the inspiration of Dr. Tilney) to take up the study of the curious midbrain structure. The work consisted of microscopic study and drawings made of the substantia nigra from enlarged projections of the epidiascope. Serial sections (Weigert stain) were studied from about the middle of the pons, through the peduncles, hypothalamic region and up to the thalamus. While the actual cell structure could not, of course, be studied, the cell grouping, etc., could be microscopically investigated. The series consisted of a human (a case of Wilson's disease), a gorilla, a chimpanzee (troglodytes niger), an orang-outang (simia satyrus), a gibbon (hylobates hoolock), a South American monkey (mycetes seniculus), a baboon (cynocephalus), the macacus rhesus, a prosimian-lemur—and the hapale jacchus. In all of these the substantia nigra was found to be neither discrete nor sharply delimited and the whole gray structure merged into numerous other gray structures from which ordinary description is wont strictly to differentiate it. The substantia seems to be continuous, caudad with the pontine nuclei, tegmentally with the gray reticular substance, laterally with the geniculate body and cephalad with the hypothalamic region. While numerous fibers seem to enter the substantia nigra from adjacent structures and others seem to emerge from it to them (a number of bundles of fibers are seen to traverse it), one cannot follow out with any degree of definiteness the various connections. Obviously more definite information could be derived from a study with methods of degeneration, but the few facts which have been gleaned justify the assumption that the substantia nigra is a somewhat ill-defined structure which stands in connection with and is but a differentiated part of the reticular gray substance.

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* Based on Professor Tilney's collection of serial sections in the laboratory of the Department of Neurology, Columbia University, New York.

THE PIGMENT

Although the substantia nigra exists in all the mammals it is pigmented only in man. The dark brown pigment is found in the cells, one third to one half of whose bodies are filled by it (Obersteiner). The pigment appears only after the third year of life, but most of the cells are still unpigmented at that age. Between five and six years the pigmented cells predominate and at about eighteen the pigment deposit has attained its maximum (Pilcz); after that it remains constant. The pigment deposit gives no fat or iron reaction (Obersteiner). The histochemical reactions point to its being a melanin similar to that found in the retina and skin, and probably is the result of the splitting of albumin by proteolytic ferments (Bauer).

There are other pigmented structures in the central nervous system, such as the substantia ferruginea (locus coeruleus), but there the cells differ in that they are rounded and vesicular in appearance. A substantia ferruginea superior is mentioned in the nuclei of the cerebellum (the superior median nucleus, nucleus tecti or fastigii). A third pigmented structure, the substantia ferruginea ventralis, is said to exist ventrally to the posterior longitudinal bundle at about the crossing of the fillet. This consists of scattered, dark pigmented ganglionary cells partly dispersed in the crossing of the fillet (Obersteiner). In animals, in contrast with the substantia nigra, the cells of the substantia ferruginea are pigmented throughout.

THE CELLS

Some of the cells of the substantia nigra have been been described as spindle shaped and others as pyramidal. Mingazzini, who studied the cells in mammals and the human fetus, states they are pyramidal, the base facing the pes pedunculi and the apex pointing toward the tegmentum. There are several groups of cells, of which the ventral are atypical. As many of the axis cylinders of the cells enter the pes the substantia nigra has been likened to a cortical ganglion such as the caudate, etc. Amaldi differed with Mingazzini in that he found similar cells in the subthalamic region, the pons and tegmentum.

Mirto is of the opinion that all the axis cylinders go toward the tegmentum and that the cells are not like the cortical pyramidal cells. Obersteiner thinks that one can distinctly recognize the pyramidal form of some of the cells; which accords with the observation of the writer. Mirto believes that the substantia nigra is formed by wandering embryonal cells, which have descended caudally from the globus pallidus through the internal capsule, and that remains of such wandering cells are to be found in the capsule of adults. The same writer also believes that the motor gray extends from the claustrum through the corpus Luysi, the substantia nigra, perhaps the red nucleus, to the pontine gray. Forel does not think that the substantia nigra belongs to the caudate and lenticular group but to the formations known as the substantia ferruginea.

Various observers (Bechterew, Jürman, Obersteiner) think that the axis cylinders travel both tegmentally and ventrally. Golgi is of the opinion that they enter the reticular substance between the two lemnisci, some of the axis cylinders dividing to go up and down, others going to the posterior commissure where they cannot be followed any further. Some are said to cross. Bauer thinks that the cells of the substantia nigra are similar to those in the corpus striatum with this difference, that there is greater abundance of glia in the former. In his opinion the substantia nigra more nearly resembles the anterior horns.

The latter author describes various

groups of cells in the human substantia nigra: a lateral group which is surrounded dorsally by fibers which go to the tegmentum (fibrae efferentes tecti of Marburg). Some of these fibers, Bauer thinks, are the fibrae subthalamicae and others just fibrae efferentes substantiae nigrae. A still more lateral group is situated near the periphery and subsequently is bordered by the internal geniculate body. There is an intermediate group, a median group, and, finally, a paramedian which is situated in the fossa interpeduncularis under the crossing of Forel. The last group of cells because of their pigmented appearance belong to the substantia nigra and not to the ganglion interpeduncularis.

In some of the simians the cell groups are similar to those in man, in others the paramedian group is lacking and in still others the lateral group is poorly developed. In many animals the cells spread into the tegmentum and can be seen in the red nucleus, lemniscus and substantia reticularis. In some of the rodentia the substantia nigra is kidney shaped, with the hilus facing dorsally. In the proboscidia the substantia nigra is split up into ganglia and is not homogeneous.

FIBER CONNECTIONS

Both Meynert and Obersteiner have described a region of fine, reticulated, braided bundles of fibers between the substantia nigra and the pes pedunculi, which are called the peduncle of the substantia nigra or the stratum intermedium dorsale. The fibers stand in relation with the cells of the substantia nigra. Most authors agree that fibers from the pes pedunculi enter the substantia nigra and axis cylinders from the cells of the substantia enter the pes. This has been universally observed in all the sections studied by the writer. Bechterew, Witkowski, Kam and Maheim showed connections of the substantia nigra with the caudate nucleus by means of secondary atrophy and degeneration. Wernicke believes that lenticular fibers enter the substantia nigra. In some instances the writer has observed whole bundles of fibers from the cortico-spinal and corticopontine regions entering the substantia, particularly laterally. Obersteiner speaks of fibers from the putamen entering the substantia nigra. The stratum intermedium is said to connect the caudate and the substantia.

According to Muratow, as shown by secondary degeneration and atrophy following the removal of the motor cortical area, the latter sends fibers to the substantia nigra. Von Monakow deduces from pathological studies and from experimental removal of the frontal cortices in new-born animals that the substantia nigra has connections with the third frontal convolutions, the insula and operculum. Rossolimo found both microscopic and macroscopic degeneration of the substantia nigra following embolic softening of the frontal, temporal and central convolutions. Maheim observed degeneration of fibers and cells and disappearance of pigment, mostly laterally, in the substantia nigra, due to a primary focus of degeneration in the region supplied by the arteria fossae sylvii. In case of cystic degeneration involving the posterior part of the thalamus, the tegmentum and the internal capsule, the substantia nigra showed degeneration in the outer third. Bechterew found degeneration in the middle third as the result of a focus in the anterior part of the internal capsule. From studies on degenerations Dejerine concluded that the central convolution sends fibers to the substantia nigra, some of them ending in it while others continue caudally in the fillet and further down unite with the pyramids. In a case of Widmer, in a longstanding circumscribed affection of the operculum, third frontal, the first temporal convolution and the putamen, the substantia nigra was secondarily degenerated.

Obersteiner believes that the substantia nigra has connection with the spinal cord. Bechterew believes the substantia has connections with the fillet and pes caudad, the former having centripetal and the latter centrifugal fibers. In a case which showed degeneration of the substantia nigra Werdnig found besides degeneration in the peduncle also ascending and descending degeneration in the lemniscus. Kölliker described fibers which entered the median lemniscus and others which penetrated it only to be lost in the tegmentum. Tschermak, Jürman and Bechterew think that there is no doubt about the connections between the substantia and the posterior white columns of the cord by means of fibers from the nuclei gracilis and cuneatus and the median fillet. Kölliker speaks of fiber connections between the internal geniculate ganglion and the substantia nigra. From the writer's own observation there seems to be no doubt of this. Indeed in many sections of most of the series, in man and the simians, one could not distinguish between or delimit the substantia and the geniculate. Bechterew speaks of connections between the lateral part of the substantia and the superior corpora quadrigemina, and Marburg mentions those fibers as running together in the tectospinal tract and in the median lemniscus to end in the substantia nigra.

PHYSIOLOGICAL INVESTIGATIONS

Economo extirpated a very minute part of the frontal convolution, just anteriorly and below the leg area, a part which on stimulation resulted in movements of chewing and swallowing, and he found degeneration in the internal capsule, in the pes, in the field of the cranial nerve fibers, in the stratum intermedium and in the fibers which entered the middle portion of the substantia nigra. The operculum has centers for chewing and swallowing, and the speech center in the third frontal convolution has to do with respiration. Trapesnikow caused circumscribed destruction of the swallowing center in the dog and degeneration resulted in the pes pedunculi, fillet and substantia nigra. Experiments of Jürman showed that destruction of the breathing center in the frontal lobe caused degeneration in the mesial part of the substantia nigra, while destruction of the swallowing center led to degeneration of the lateral part. Economo maintains that there is a center in the substantia which aids in the chewing movements and those dealing with the intake of food On extirpation of those parts of the cortex, which subsequently revealed degeneration in the substatia nigra, there resulted interference with the ability to swallow, but as the animals with the ablated cortices regained control after a shorter or longer period, Economo concluded that the center in the substantia nigra is independent of cortical control. Brissaud thought that the substantia nigra had something to do with Parkinson's disease, as he found a solitary tubercle in it in a patient who died from paralysis agitans.

By means of specially constructed electrodes Jürman stimulated directly the lateral portion of the substantia nigra and repeated swallowing movements resulted; fewer movements followed stimulation of the central portion and fewest that of the mesial. Besides the swallowing movements there also occurred contractions of the anterior neck muscles, the orbicularis oris and the palatal muscles. Stimulation of the lateral portion tended to check respiration, mainly in expiration, while stimulation of the mesial portion stimulated respiration. In two animals in whom he succeeded in producing isolated destruction of the substantia nigra (the animals lived two and three days respectively) no motor or sensory disturbances were noticed.

Epidemic encephalitis possibly furnished clinical evidence of the function of the substantia nigra. One occasionally encountered peculiar champing movements during the course of disease in patients who showed symptoms of midbrain involvement, which may be interpreted as the result of an irritative lesion in the substantia nigra. Many of the movements which have been observed were distinctly of the chewing and swallowing type, although most frequently one saw an up and down, champing movement. The epidemic of encephalitis may, therefore, be said to have thrown some light on the function of the midbrain, the site which was so frequently attacked by the virus.

DISCUSSION

From a study of the serial sections of the substantia nigra in man, in seven simians and two other animals and from a review of the comparative anatomical studies of Bauer it appears that the structure is about the same in all mammals, although it is best developed in the human. It is not at all well defined and microscopically one cannot delimit it from the tegmentum, internal geniculate, the pontine nuclei or the hypothalamic region. In some of the animals it either goes down to a lower level or reaches a higher one than in others. According to Bauer it contains four groups of cells. In some of the apes the intermediate group extends into the lemniscus and tegmentum. In man (as well as in the cebus and orang) there is a paramedian group.

Like other gray matter the substantia nigra is densely interlaced with white fibers and traversed by numerous bundles of fibers. Numerous fibers enter it from the pes pedunculi, from the tegmentum, internal geniculate body and other adjoining structures, and it probably sends fibers to all these structures, although it is frequently difficult to point out the direction of the fibers. The fibers which lose themselves in the central gray are called fibrae efferentes; more cephalad, there are the fibrae efferentes tecti of Marburg or the fibrae subthalamicae substantiae nigrae. Fibers of the pedunculus corporis mammillaris traverse the substantia nigra mesially.

The substantia nigra is found in all mammals but is said not to exist in lower vertebrates. According to Ziehen it is poorly defined in the monotremes. No studies of it have been made in birds. The avian nucleus entopeduncularis described by Edinger is said to be the homologue of the substantia nigra in mammals. Bauer believes that the substantia nigra was originally an undifferentiated structure and that its cells have developed from the central gray, from which they moved ventrally; hence the reason why some of the cells are found in the lemniscus and reticular substance. This is contrary to Mirto's view that the substantia developed from the lenticular nucleus. As the cells were differentiated from the tegmental gray they moved toward the pes fibers with which the substantia nigra stands in intimate connection. Further, according to Bauer, with the development of the basal ganglia and cortex, the substantia nigra received connections from them. It stands as intermediary, together with other tegmental structures with which it has connections, for the purpose of coordination.

"Just as the numerous cells grouped throughout the brain-stem and conceived by Edinger as the nucleus motorius tegmenti are under the influence of the cerebellum so the cells of the substantia nigra are for the purpose of coordination under the influence of the cortex. The cells carry impulses from cerebral centers, from the cortex or corpus striatum, directly or through association cells of the tegmentum, to various efferent cell complexes. The coordination of chewing and swallowing is only one of the numerous associational and coordinative functions of the substantia nigra."

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[For a complete bibliography see Bauer's article on the substantia nigra.]

EXTRA-MEDULLARY FIBRONEUROMA MAINLY INVOLVING THE POSTERIOR COLUMNS OF THE CORD

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Patient, Mrs. A. R., aged thirty-nine, seamstress.

Chief Complaint.—Numbness of the left leg for two years; numbness of the right foot for three months.

Family History.—The father and mother are dead; the father died at fifty-two of acute intestinal obstruction, the mother at sixty, of cancer of the liver. The patient has five brothers and three sisters all living and well. There is no history of tuberculosis, rheumatism or gout. The grandfather died of heart disease. There has been no Bright's disease, diabetes or obesity in the family.

Personal History.—She has been married twelve years. Her husband is healthy. She has not lived with her husband for the past year. She has never been pregnant. She gets little exercise but her habits of eating and sleeping are good. She takes one or two cups of coffee a day, no tea, alcohol, drugs or medicines. The patient is a seamstress working in her own home. The best weight was 218 pounds three years ago, the average weight is 203 pounds, as is also the present weight.

Previous Health and Illnesses.—Patient was always healthy and robust. She does not remember any childhood illnesses except frequent attacks of tonsillitis, and has had no illnesses up to the present time. Six years ago both ovaries were removed for cysts.

Head.—No headaches. Eyesight good, the patient wears glasses. She has colds in the head frequently; no epistaxis. Ears have always been normal—no discharge or tinnitus. Teeth have always been in good condition.

Present Illness.—Two years ago she commenced to have a sensation of numbness in the third and fourth toes of the left foot. This gradually spread to the ankle and knee. A few weeks later she noticed that the knee was getting stiff and that she would stumble when getting on and off street cars. The ankle began to turn outward and this made walking more difficult and increased the tendency to fall. About a year ago for a period of two weeks, when she was perfectly quiet at night, the whole limb would suddenly be drawn up, but would be relaxed again in the course of a few seconds. This was only when she was lying down, but never when asleep. At this time she had severe pain in the back of the left buttock. This was dull and boring in character and pretty constantly present for a period of a few weeks. It was most marked at night.

One year ago she commenced to have trouble with micturition. At first she noted that there was considerable delay in starting urination. At times she was unable to void though the bladder felt distended. This has continued to the present. Now at times, when voiding, the urine will stop suddenly for a few seconds and then flow again freely. When it ceases suddenly in this way there is a sensation of "jerking" on the inner side of the left calf. There has never been any incontinence. Her bowel control is normal unless she has taken a cathartic and the movements are loose.

At present the numbness has extended nearly to the hip and the whole leg is stiff, necessitating walking with a stick. A few months ago the numbness commenced in the right foot and has gradually spread to the right knee.

PHYSICAL EXAMINATION, NOV. 19, 1920

General Appearance.—A well developed, slightly obese, healthy looking, middle-aged woman lying comfortably in bed. Her color is good, her skin free from rashes or eruptions. The hair is normal in amount and distribution. Mentally she is bright and does not appear to be of neurotic type. There is no speech defect. Weight is 196 pounds.

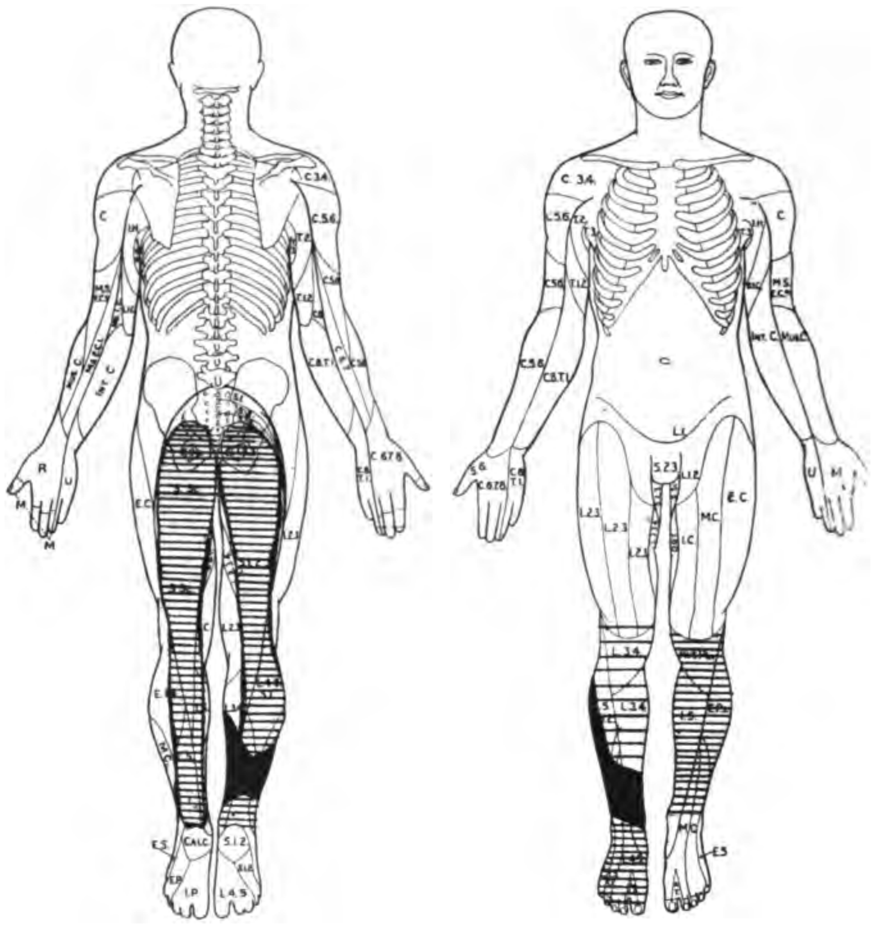

Fig. 1. Disturbance of Temperature Sense.

Head.—Is well formed; there is no mastoid or sinus tenderness.

Eyes.—The ocular movements are normal. Conjunctivae and sclerae show no abnormality. Error of refraction is corrected by glasses.

Nose and Ears appear normal; no discharge.

Mouth.—The mucous membranes are normal. The tongue is not coated; it protrudes in the midline and is free from any tremor. The gums and teeth are in good condition, though there has been considerable dental repair with many fillings and a few extractions. The tonsils are enlarged and cryptic but not inflamed. The pharynx is normal.

Neck.—No abnormal masses or pulsations. The thyroid is not enlarged. The neck is not stiff.

Thorax.—It is well developed. The respiratory movements are equal on both sides with good expansion. The breasts are well developed and contain no abnormal masses.

Heart.—The apical impulse is not visible or palpable; and there is no enlargement to right or left. The sounds are of normal quality. There are no murmurs or other adventitious sounds. The pulses are equal, regular, low tension, and the vessels soft.

Blood pressure—systolic 125, diastolic 88.

Lungs.—Percussion is normal throughout; expansion at bases is good. The breath and voice sounds are normal. There are no râles.

Abdomen is flat, soft and slightly pendulous. Below the umbilicus in the midline there is an operative scar four inches in length, the wound is firmly healed. The liver edge is palpable on deep inspiration. The spleen and kidneys are not felt. There are no abnormal masses, and no signs of fluid.

Pelvic Examination.—Vaginal: no tenderness, cervix small, points forward. No masses. Rectal: negative.

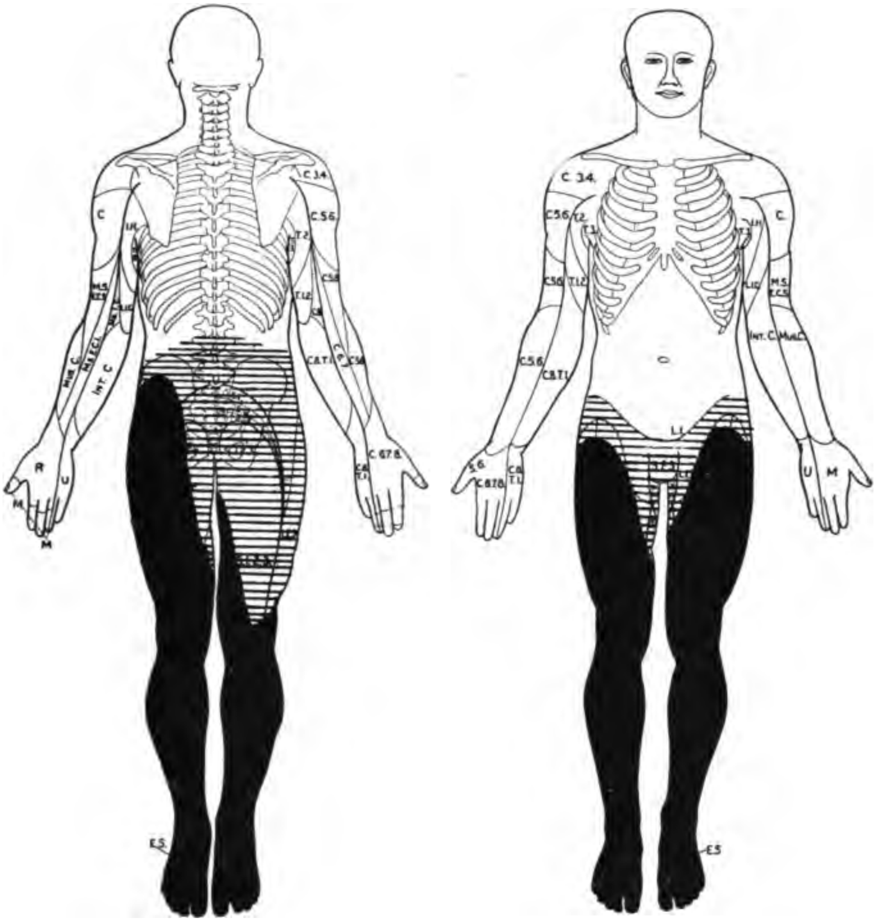

Fig. 2. Loss and Diminution of Vibration Sense.

NEUROLOGICAL EXAMINATION NOV. 20, 1920

General Appearance.—Patient is a rather obese, normally developed woman who does not appear acutely ill or in discomfort. There is no bony deformity noted. On standing there is a tendency of the left foot to become everted, and on lying in bed with the feet relaxed, the foot constantly assumes a partially everted position.

Gait.—On walking forward the left foot seems stiff and spastic. It has a tendency to become everted and the toes to turn inward, the heel on this side is not lifted high from the floor. She complains that on walking a short distance she becomes quite fatigued.

Coordination.—Equilibratory: Standing with the feet together, with the eyes open, there is moderate swaying. This is much more marked when the eyes are closed. She is unable to stand on either foot alone with the eyes either open or closed.

Non-equilibratory: The finger-to-nose test and pointing and past-pointing in both horizontal and vertical planes with each arm, are performed with precision. The heel-to-knee test is carried out fairly accurately with the right leg, though there is some wavering and imprecision. She is unable to flex the left thigh and leg sufficiently to place the heel on the right knee.

Diadochokinesis: Rapid pronation, supination, flexion and extension of the forearms and arms are normally performed. She is able to flex and extend the right leg with fair rapidity, but is unable to carry out this movement in the left leg, mainly because of the increased spasticity.

Skilled Acts.—Voice and speech are normal and handwriting is unaffected.

Reflexes:

DEEP	*Right*	*Left*
Jaw	2	2
Pectoral	2	2
Biceps	3	2
Triceps	2	2
Radial	2	2
Ulnar	2	2
Supra-patellar	2	3
Patellar	2	3
Achilles	2	2

SUPERFICIAL		
Supra-umbilical	Present	Present
Suprapubic	Present	Present
Upper and lower lateral abdominal	Present	Present
Plantar flexion	Absent	Absent
Babinski	Present	Present
Chaddock	Absent	Present
Oppenheim	Absent	Absent
Gordon	Absent	Absent
Schäfer	Absent	Absent

0 = Absent; 1 = Slight; 2 = Active; 3 = Very active; 4 = With transient clonus; 5 = with permanent clonus.

Abnormal Involuntary Movements.—None noted.

Muscle Strength.—There is evident diminution in all the movements of the left thigh, calf and foot. Otherwise the muscle strength seems to be normal.

Muscle Status.—Normal.

Abnormal Associated Movements.—Not tested.

Nerve Status.—No observation.

General Sensory.—*Touch:* Perception of cotton is diminished over both legs but is absent only on the soles of the feet and in a small area about two inches in diameter over the anterior portion of the left ankle. There is no sharp dividing line between the abnormal and normal sensation, but sensibility starts to be diminished on the left side about the middle of the thigh, and on the right side below the knee.

Discrimination between sharp and dull is normal.

Pain sense is diminished over the same areas, but is nowhere absent. Pain on deep pressure is possibly less marked in the right leg than in the left.

There are distinct points of tenderness over the spines of the second and third lumbar vertebrae, especially over the third.

Two-point Recognition.—There is considerable disturbance in the recognition of two points over both legs. This is more marked on the left side than on the right. On the outer side of the left calf the points must be separated 22 cm. before they are perceived as two. On the corresponding portion of the right calf they must be separated 12 cm. Over the left thigh the distance is 8½ to 12 cm.; over the right thigh, 7 to 8 cm.; while over the abdomen the distance is 3 to 7 cm.

Temperature.—It is difficult to outline the diminution in temperature sense, but it seems to be more marked on the right side.

There is a band about the right ankle three inches in diameter, and an area on the outer side of the right calf where heat and cold are not recognized at all. The sensitivity is diminished over the entire right leg below the knee. On the left side there is some diminution of discrimination over the entire leg between the ankle and knee, but it is nowhere absent. Posteriorly there is some loss of discrimination on each side over the middle of the thigh as high as the buttock.

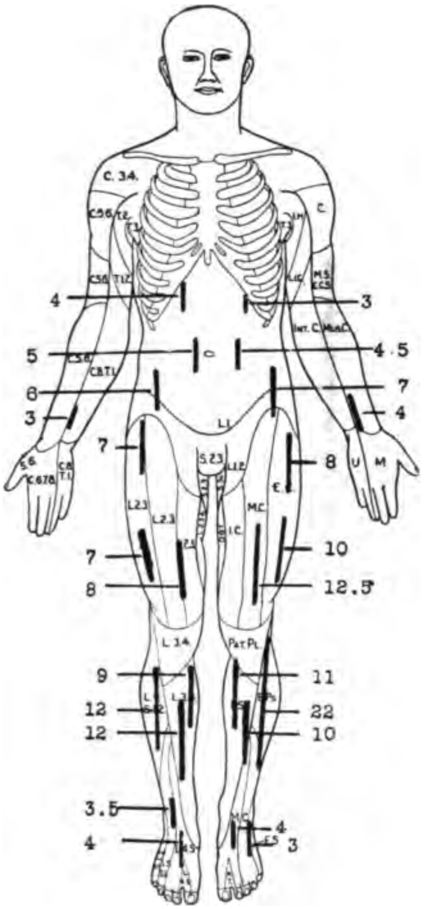

Fig. 3. Compass Test. Distance (in centimeters) points must be separated to be recognized as two.

Vibratory.—The vibrations of a tuning fork are lost over the entire right leg with the exception of a small portion on the inner side of the thigh as high as the crest of the ilium, where the vibrations are faintly perceived. On the left side vibratory sense is absent below the knee. On applying the tuning fork to the spines of the vertebrae, there is distinct diminution below the second lumbar. Above this and over all the dorsal vertebrae and ribs vibration is equally well recognized.

Muscle Tendon Sense.—The patient is absolutely unable to tell the position of the toes on the left side when they are markedly flexed or extended. Slight movements of the left ankle and knee are also unperceived. There is some disturbance of recognition of position in the right toes, but it is not nearly as marked as on the left side.

Stereognosis.—Familiar articles are recognized in either hand.

Cranial Nerves and Head. *Olfactory Nerve and Nose.*—The sense of smell is normal in each nostril.

Optic and Oculomotor Apparatus.—Vision seems normal. Perimetry, tested with the hand, shows no contraction of any field. Eyegrounds are normal. The pupils are moderately dilated, circular, regular and equal. Both react actively to light and on accommodation. The palpebral fissures are equal and normal. There is no exophthalmos, enophthalmos or ptosis. Strabismus and nystagmus are absent. There is no limitation of the conjugated eye movements.

Acoustic Nerve and Ear.—Hearing is normal on each side. Air conduction is greater than bone.

Trigeminus Nerve and Mouth.—The corneal reflex is present on each side. The movements of the jaw are free and equal.

Facial Nerve and Face.—No weakness of either side can be determined.

Glossopharyngeus and Vagus Nerves, Pharynx and Larynx.—The uvula is in the median plane. Both palatal reflexes and the pharyngeal reflex are present. Swallowing is normal.

Spinal Accessory Nerve.—Normal.

Hypoglossus Nerve and Tongue.—The tongue is protruded mesially and moves normally in all directions.

LABORATORY EXAMINATIONS

Blood Wassermann, November 17, 1920

Alcohol antigen	Negative
Cholesterin antigen	Negative

Spinal Fluid

Amount	15 c.c.
Appearance	Clear
Pressure	Normal
Cells	10 lymphocytes
Globulin	Negative
Wassermann	
Alcohol	Negative
Cholesterin	Negative

Gold curve 4.5, 4.5, 4.5, 3, 3, 3, 2, 1, 0. 0.

X-Ray Examination of Lumbar Spine

No indication of disease in the lumbar vertebrae except for a slight lipping in the lumbosacral joint.

Urine November 22, 1920

Color	Amber
Reaction	Acid
Specific gravity	1.018
Turbidity	None
Albumin	None
Glucose	None
Microscopic examination	Negative

SUMMARY AND CONCLUSIONS

The history is that of a gradually progressive lesion of the cord of two years' duration. For the first year and a half the symptoms were those of a hemiparaplegia, and it is only during the past few months that evidences of involvement of both sides of the cord have developed. At present the motor symptoms are mainly confined to the left side and consist of weakness of the muscles of the thigh, calf and foot, hyperactive knee and ankle reflexes and a positive Babinski. The Babinski reflex is present also on the right side, though there is no definite muscular weakness demonstrable in this limb.

The sensory disturbances are mainly confined to the posterior column. There is hypaesthesia over the left leg below the center of the thigh, and of the right leg below the knee. The sensations of pain and temperature are not normal, the disturbances being more on the right side, but not confined to areas that make them of much localizing value. Determination of position and passive movement is defective in the left toes, ankle, knee and possibly the hip, and also, but to a less extent, in the right toes. Vibration sense is lost over both legs below the crests of the ilia, except on the upper and inner surfaces of the thighs. It is diminished below the groin. Posteriorly, the loss is not as extensive, being only partial on the upper three-fourths of the right thigh and over the inner surface of the left thigh. However, there is a definite diminution everywhere below the spine of the second lumbar vertebra. On the vertebral spines and the soft tissues above, the vibrations are everywhere very distinct. The disturbance of vibratory sense thus gives the most definite evidence as to the level of the lesion, and places it at about the second lumbar segment.

The progressive development of a localized lesion of the cord would suggest a neoplasm or possibly a gummatous formation. The latter is ruled out by the negative blood and spinal fluid findings. Multiple sclerosis is unlikely in view of the fact that the history indicates a single lesion involving first one half of the cord and gradually spreading to the other. The negative x-ray and absence of root symptoms would indicate an intramedullary lesion. The tentative diagnosis is therefore an intramedullary tumor of the cord at approximately the level of the second lumbar segment.

DISCUSSSION

Dr. Strong. It seems to me rather hard to localize this lesion with completely consistent results. The pain and temperature disturbance below the level of the lesion seems to show that we must be dealing with a gray lesion and not a tract lesion. On the other hand the loss of position, movement and vibratory senses points to the posterior columns.

I agree with Dr. Howe in regard to the location of the lesion, although it seems unusual that the pain and temperature disturbances are on the opposite side. When we get a bilateral lesion like that it is rather difficult to decide on the exact location. I believe that the major lesion is on the left, but extending over to the other side.

Dr. Wechsler. It strikes me that it is not quite in the cord itself. If so we would have greater disturbance of the bladder and rectal control. I believe the tumor is extra-medullary.

This shows a little bit of a Brown-Séquard symptom, such as we are apt to see clinically, with loss of pain and temperature sense.

As to the location of the lesion, if it is at the second lumbar segment, we ought not to have an increased knee jerk. The sensory symptoms—at least the vibratory, go to the 12th dorsal or the first lumbar. I should say that the lesion is two or three segments higher, about the 12th dorsal.

As to treatment, it seems to me that an exploratory operation is indicated.

Dr. Casamajor. The points that Dr. Howe made are pretty carefully brought out. The lesion can hardly be doubted. As Dr. Strong pointed out, I think it is in the gray. And it seems to me that we are dealing with an intra- and not an extra-medullary lesion.

I think that the question of syringomyelia should be considered here. The patient is a bit old for a beginning syringomyelia but nevertheless not too old. The presence of this fairly rapidly growing lesion beginning in the cord and extending into the posterior columns might as easily be a syringomyelia. Syringomyelias start in that posterior primitive cell spur and generally enlarge posteriorly, and then as a rule turn, but keeping a good part of the way in the posterior column.

There seems to be an absence of the pressure signs in this case that one would expect, especially an absence of globulin in the spinal fluid and also an absence of the yellow color in the fluid. So that we must be careful in making a diagnosis of neoplasm, especially in view of operation. As far as treatment of the case is concerned I think an operation is indicated. Surely an exploratory operation can do no harm if it is syringomyelia. If it is an intramedullary lesion, it might be small enough—provided it be an endothelioma—to be removed. In any event it is worth a trial.

I think the patient should be advised to have an operation. The prognosis should be guarded for the possibility of syringomyelia must be considered. The normal spinal fluid and the normal serology exclude syphilis. But the gold curve would not necessarily exclude syringomyelia.

Dr. Tilney. Tingling in the left great toe is certainly not an intramedullary symptom in my opinion. My first interpretation would be that it was an irritative lesion, probably extramedullary or root. The stiffness and weakness in the left leg could easily enough fall into that same group. The occurrence of the dysuria looks like an irritative change, I should say an irritative cord disturbance. Pain in the buttock, even transitory pain, to my mind is one of the most important symptoms which we have in cord extramedullary lesions. Dr. Riley and I have had a case under observation for over a year which we had interpretated in various ways. There was a history of pain in the left shoulder for a short time on two occasions. At operation a cord tumor was found. That pain to my mind indicates an irritative root lesion.

Now as to the dysuria and the type of dysuria which the patient has. Apparently when the bladder becomes distended with urine the patient has difficulty in the control of the sphincter, and either rapidly expels the urine without control or else has retention, or, when it is passed, has a sensation of pain. In my experience that is not the kind of thing we get from intramedullary tumors, but it is seen in low extramedullary cord tumors and therefore I think it fits into that picture more than into any other.

Numbness in the left leg seems to indicate an extension of the lesion, passing more into a destructive phase and producing a sensory loss subjectively. This extension also shows that the lesion is a growing one. Numbness of the right foot shows a still further extension of the tumor.

At the present time the patient has an ataxic hemiparaplegia greater on the left than on the right. We know that the left pryamidal tract has been compressed, is more or less out of commission, and that the right is becoming impaired because we have upper motor neurone symptoms on both sides.

That would be an unusual thing in an intramedullary lesion. It might occur, but it seems to me more of a destructive process, crossing from the left until it has involved the right.

The sensory changes do not resemble those of syringomyelia or multiple sclerosis.

The condition seems to me to be the result of an irritative disturbance which gradually extended until it became a destructive one; and from the fact that there were root signs, it is more probably extra- than intra-medullary.

As to localization, there can be no doubt that it is a cord lesion on account of the sensory and root symptoms.

In making a tentative diagnosis I would put extra-medullary neoplasm first. I would consider an extra-medullary one, but put it lower on the list, with syringomyelia still lower, and multiple sclerosis last of all.

Finally, as to the treatment: A case like this unquestionably deserves an exploration, as a result of which I should expect to find a cord tumor at about the level of the first lumbar vertebra.

DR. OSNATO. Would Dr. Tilney give any localizing importance to the nature of the bladder disturbance?

DR. TILNEY. Yes. I think that a lower lumbosacral involvement would give just this type of dysuria.

DR. STRONG. One point I would like to raise is—is the hemiparaplegia really ataxic or the result of weakness?

Another point; I do not think an extra-medullary location explains the dissociation of sensory changes.

DR. STOOKEY. Does Dr. Tilney consider the tingling indicative of a root lesion? If so, we ought to call the point of tingling the initial point at which the tumor began, and then the level which supplies the left great toe would be the site of the beginning of the lesion.

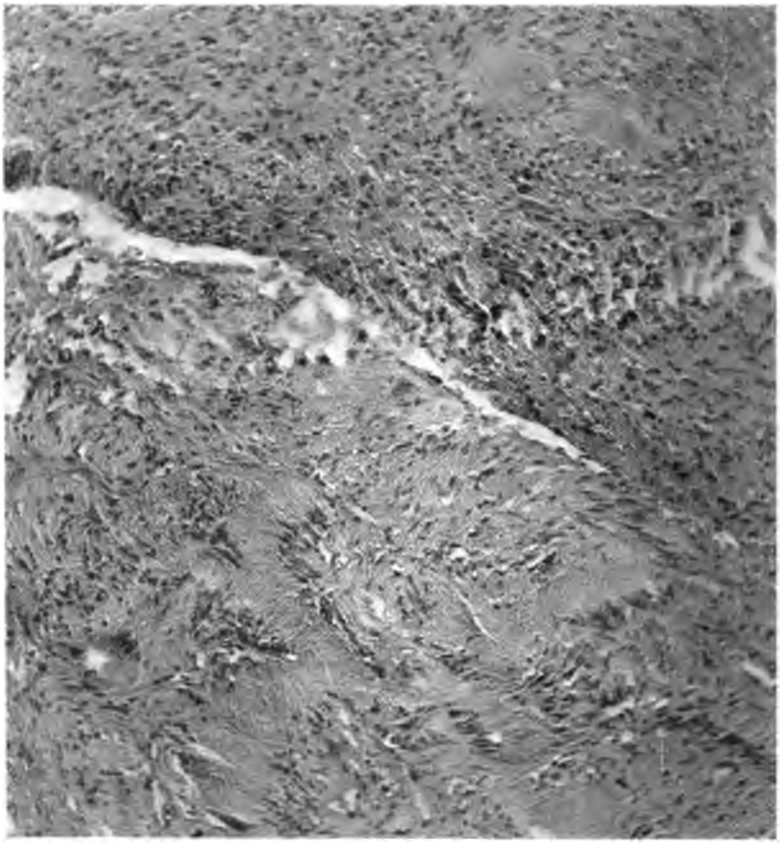

FIG. 4. SECTION OF TUMOR SHOWING HISTOLOGICAL CHARACTER OF GROWTH—FIBRONEUROMA.

Dr. Tilney. From high cervical lesions we sometimes get low cervical symptoms. For example, in the case I cited, there was atrophy in the hands with a tumor confined to the fifth cervical segment of the cord—way below the site of the tumor.

OPERATION

January 11, 1921. Laminectomy by Dr. Lambert. A tumor was found on the dorsal aspect of the cord at the level of the second and third lumbar segments. The tumor was about 1½ cm. in length and nearly as broad as the cord. It was not adherent to the dura at any point. It was dissected free from the cord and seemed definitely outlined and encapsulated. The tumor was firm, homogeneous and sectioned like fibrous tissue.

PATHOLOGICAL EXAMINATION OF TUMOR TISSUE

The section was composed of two morphologically different types of cells. One portion had the appearance of fibrous tissue and occurred in bands, while the other was homogeneous, loosely connected tissue, and formed the main bulk of the tumor. The fibrous bands coursed irregularly through the section. In some areas they formed spiral whirls, while in others they were interlaced in a reticular manner, forming a coarse network. The cells forming these bands were long and spindle-shaped. The nuclei were long and narrow and in some areas were arranged in parallel rows, the portion of the fibers between these collections of nuclei having a hyalinized appearance. The principal tissue was made up of cells with rounded nuclei and indistinct cell outlines. These cells were loosely connected together and irregularly disposed.

The appearance of this tumor was very similar to those arising from the acoustic nerve.

Diagnosis.—Fibroneuroma.

DISCRETE THERMAL CHANGES IN SPINAL SYPHILIS *

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The dissociation in thermal sensibility sometimes seen in intra-medullary spinal lesions, is a matter of great interest to us, because up to the present time we have no established anatomical explanation of the phenomenon. Our clinical experience assures us however that after peripheral fibers have become recombined and redistributed within the cord substance, those carrying heat impulses are discretely separated from those carrying cold. And there is abundant evidence that the pain elements are likewise discretely placed within the spinothalamic tract. The work of Head emphasizes, upon theoretical grounds, the establishment within the cord of secondary paths for the transmission of specific afferent stimuli; but the close anatomical association of the three forms of protopathic sensibility within the spinothalamic pathway makes it difficult for us to understand the escape of one or another form when the lesion is manifestly a diffuse one.

Such a problem is encountered in the following case, which I present from the standpoint of differential diagnosis, as well as to record these rather uncommon sensory disturbances.

PATIENT.—Frank B.; married; German born; aged thirty-eight; occupation, "tester."

Present Illness. Complaints.—1. Weakness in both legs, the right being more affected than the left. 2. Great difficulty in walking. 3. Pain in back. 4. Painful micturition. 5. Constipation.

Date of first symptom.—Three and a half years ago. Onset, gradual; progress, worse.

The patient had not had any serious illness up to the onset of his present trouble. He had a so-called gonorrhea ten years ago, with "pimples" on his thighs and abdomen which lasted two or three weeks. Six years ago he had headache that was constant for about three weeks, which gradually subsided under medication. There has never been any vomiting.

With reference to his venereal infection he says that he has had two blood Wassermann tests and two spinal fluid examinations that were negative. However, he has had other blood examinations, the results of which he does not know. A physician gave him thirty mercury injections and tablets by mouth.

Four years ago he and a helper carried a kitchen range up two flights of stairs, and he recalls that this effort was a great strain upon him, but he did not note any untoward results.

In January, 1917, while at work in a very cold warehouse, he noticed a severe pain in his urethra when he attempted to pass water. This pain became progressively worse until he went to a physician whose medicine he thinks relieved him for a time. During the period of January to April of that year he had also noted a progressively increasing tiredness in his legs, until the first of April, when it became almost impossible for him to move the right leg. As far as he can recall the left leg was not affected beyond being easily fatigued. He believes that in the past three years the right leg has become more useful. The left leg first became troublesome in August, 1920, the complaint being a sensation of "pins and needles" when he stepped upon the left foot.

The patient has indulged excessively in coffee and alcohol. He was formerly employed in a wine room, and drank constantly up to four years ago. Since the onset of his illness he has not taken alcohol in any form. His sexual power has diminished considerably. His bowels are very much constipated. Micturition is painful to him, frequent, and extremely urgent. At times he is incontinent if he does not reach a toilet in time. He has nocturia once or twice.

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*Presented at a clinical conference at the Neurological Institute, from the service of the Fourth Division.

The family history is entirely negative.

NEUROLOGICAL EXAMINATION

Abnormal Attitudes and Deformities.—Patient walks with a cane, bent slightly forward.

Gait.—Typically spastic gait, right leg being more involved than the left. Automatic associated movements of arms, head and trunk normal.

Coordination.—There is no equilibratory disturbance. There is apparently no non-equilibratory ataxia, although the patient's legs are so hypertonic that it is impossible to perform the heel-to-knee tests. The arms are normal. There is no adiadochokinesis.

Skilled Acts.—Handwriting is normal. Speech is normal.

Abnormal Involuntary Movements.—There is some tremor of the outstretched hands and fingers, but it does not seem pathological.

Reflexes:

DEEP	*Left*	*Right*
Jaw	not elicited	not elicited
Pectoral	not elicited	not elicited
Biceps	2	2
Triceps	2	2
Radial	2	2
Supratellar	3	4
Ankle clonus	0	permanent
Patellar	3	4
Achilles	4	5
SUPERFICIAL		
Supra-umbilical	0	0
Suprapubic	0	0
Upper lateral abdominal	0	0
Lower lateral abdominal	0	0
Plantar flexion	0	0
Babinski	1	1
Fanning	0	0
Chaddock	1	1
Oppenheim	1	1
Gordon	0	0
Schäfer	0?	0?

1 = present; 2 = active; 3 = very active; 4 = very active with transitory clonus; 5 = with permanent clonus; 0 = absent.

Muscle Strength.—Arm and trunk groups are normal. Legs not satisfactorily tested because of hypertonicity.

Muscle Status.—No atrophy or hypertrophy noted. Marked hypertonicity of lower extremities. Arms normal.

GENERAL SENSORY.—The sensory examinations were made on many occasions, and identical results were obtained.

Touch.—There is a very questionable diminution over the posterior portions of the leg area, right and left, but everywhere else touch is normal.

Pain.—There is no deviation from the normal in any part of the body. Deep and superficial pain sense are alike unaffected.

Temperature.—Here very striking results are obtained. Cool and ice cold stimuli are interpreted correctly all over the body. The lower extremities are examined repeatedly, and the findings are constantly the same.

On the other hand, warm and hot stimuli are interpreted almost invariably as "cold" in the lower extremities. The marked disturbance reaches as high as the iliac crest line anteriorly and to the mid-gluteal region posteriorly. Above these lines, the sensation is absolutely normal.

Vibration.—Vibratory sense is normal in all parts of the body.

Muscle Tendon.—There is no disturbance of position or joint sense in the lower extremities.

CRANIAL NERVES.—*Olfactory and nose* not examined.

Optic, oculomotor, trochlear and abducens nerves and optic apparatus: Fundi, normal. Pupils, equal, central, regular, reacting well to light and convergence. There are no limitations of movement of the eyes. No nystagmus, ptosis, exophthalmus, or enophthalmus.

Palpebral fissures equal. Patient says he has had an occasional squint since birth. (Dr. Ward Holden reports eyes negative, except for periodical convergent strabismus of long standing.)

Acoustic Nerve and Ear.—Not tested.

Trigeminus and Mouth.—Motor fifth normal. Sensory fifth normal. Several teeth need investigation.

Facial and Face.—Taste not tested. Motor seventh normal in all tests. Associated movements of face, eyes and head normal. Vasomotor status normal.

Glossopharyngeus, Vagus, Pharynx and Larynx.—Uvula rises in mid-line. Phonation normal, swallowing normal.

Spinal Accessory.—No abnormality.

Hypoglossus and Tongue.—Tongue protrudes in midline. Movement in all directions normal. Trophic status normal.

MENTAL STATUS.—Normal intelligence. Emotional state normal. Attention and memory normal.

SYSTEMIC.—Tegumentary status normal. There is a marked thickening of the skin of the sole of the left foot. Glandular status normal. Cardio-vascular: pulse, normal. Systolic pressure, 140. Heart negative. Abdomen negative. Genito-urinary (see history.)

Skeletal.—X-ray of dorsal and lumbar spine shows evidence of bony excrescence formation between the outer borders of the third and fourth lumbar vertebrae, and along the upper surface of the third lumbar vertebra.

LABORATORY TESTS.

Blood Wassermann:

Sept. 28, 1920	one plus
Oct. 1, 1920	four plus
October 5, 1920	four plus

Spinal Fluid.—Sept. 28, 1920. No cells, globulin not in excess, sugar present, Wassermann negative. Colloidal gold 11111O.

SUMMARY OF PATHOLOGICAL FINDINGS

1. Spastic paraplegia.
2. Absence of thermal sensibility for warm and hot stimuli up to the level approximating lower dorsal segments, with preservation of cool and cold sensibilty.
3. Osteo-arthritis of lumbar spine.
4. Three positive Wassermann reactions in the blood serum.

DISCUSSION

From the standpoint of symptomatology, it is essential to recount the oustanding complaints from which the patient has suffered in the past three and a half years. The dysuria with which his trouble began undoubtedly represents an organic disturbance of the cord substance. The progressive weakness of the legs represents, to my mind, a vascular degenerative process affecting chiefly the lateral pyramidal tracts. While there never were signs of intermittent claudication of the spinal type, the history is suggestive of such vascular changes. The patient has become gradually conscious of the involvement of both lower extremities. We have seen in our examination the positive signs of bilateral pyramidal tract disturbance in the cord. The frequency and urgency of micturition are explained by these signs. The "pins and needles" sensation in the left foot is probably due to the thickened skin of the sole. The pain in the back is probably accounted for by the osteo-arthritis.

We are dealing therefore with a gradually progressive disease which has thus far been limited to the lateral pyramidal tracts, with evidence also of dissociated thermal sensibility.

Primary lateral sclerosis does not present sensory symptoms. Multiple sclerosis, on the other hand, may give us this picture, but I believe the patient's age, absence of eye signs, speech disturbance and variation in symptoms, quite eliminate such a possibility after the lapse of nearly four years.

Spinal cord tumor may frequently give such a history, but I believe with such evidence of diffuse bilateral disturbance, the sensory symptoms should be much more extensive. There is no indication of a level lesion. The spinal fluid shows no xanthochromia.

Syringomyelia has in several instances shown disturbances in warm and hot perception before loss of cool and cold. As a matter of fact we find in the literature many clinical cases representing slow compression of the cord in which this dissociation has been noted during the onset. But in long-standing cases such a finding is extremely rare. I believe that in this particular instance syringomyelia may be ruled out because of extensive pyramidal involvement plus the above peculiar sensory signs with no involvement of the anterior gray or posterior columns.

The picture presented above is probably that of syphilitic degenerative processes within the cord substance. The predilection of vascular syphilis for the lower dorsal and lumbar regions is well known. The blood serology is strongly positive. The course of the disease is quite characteristic. The explanation of the sensory findings is not satisfactorily made. In such a diffuse lesion as we must have here it is most difficult to explain the significance of such a dissociation of sensation within the spinothalamic tracts, disturbance in which usually gives us not only thermal disturbances to all forms of stimulation, but also interference with pain perception. It is certain however that in this case, in spite of their close relationship, the cold and pain pathways have entirely escaped, while the fibers conveying heat impulses have been profoundly affected.

With reference to treatment, I advise intravenous neo-salvarsan, mercury by inunction, massage, and passive movements.

The prognosis in this case is not good, but with proper attention I believe the patient may reasonably expect that the progress of the disease will be checked.

THE DIAGNOSIS OF MULTIPLE SCLEROSIS IN THE ABSENCE OF THE TRIAD OF CHARCOT

By JOSHUA ROSETT, M.D.

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What is multiple sclerosis clinically? That is the particular interest attaching to this case. The symptomatology indicates a multiplicity of lesions within the central nervous system, although none of the symptoms of the classical triad of Charcot is present. The patient presents neither scanning speech, intention tremor, nor nystagmus. On the other hand, dissemination of the pathological process is manifested in many other clinical details.

The patient, a healthy-looking married woman, twenty-five years of age, came to the Vanderbilt Clinic on December 8, 1920, with the following history:

In December, 1916, six months after she had given birth to her first child, she had a mild attack of acute nephritis of a few days' duration. While she was recovering from the illness, a week after its onset, her legs became weak and stiff; and the disability increased so rapidly, that in two or three days she could neither walk nor stand. In a recumbent position, and with her eyes open, she could move her legs in any indicated direction, but she could not tell which way her legs were moved when her eyes were closed. She thinks she had lost the sense of touch in her lower limbs, as well as the sense of direction. This condition continued almost unabated for a year. At the end of that time the strength in her legs gradually returned, and with it, the sense of touch and of direction of movement. From that time on she was quite well until September, 1920, when she once more became lame. The second attack lasted for only a few weeks and was not quite as incapacitating as the first. There were periods of remission varying in duration from a few hours to a few days, when she could walk, although with difficulty; and she had no sensory disturbances. The disability gradually passed away, and for some weeks preceding her first appearance at this clinic she had been quite well.

In the anamnesis it appears that the patient had always been in very good health until the attack of nephritis and subsequent paraplegia in 1916; that she had never suffered any accidental injury and had undergone no operations; that at no time has she had any convulsions, or suffered from visual, auditory, mental or emotional disturbances; that for the last five years she has been happily married to a healthy, well-to-do workingman, during which time she has given birth to two healthy children; that she has had no miscarriages; and that her habits are normally moderate.

The neurological examination made on December 8, 1920, brought out the following facts:

There were no abnormal attitudes or deformities. Her gait, except for a barely perceptible spasticity of the right lower limb, was normal in all its phases, and so were the associated movements of the trunk, arms and head.

Equilibratory Coordination. — Standing with her feet together and her eyes open, the patient was slightly unsteady; and she swayed distinctly to the right when her eyes were closed.

Her *non-equilibratory coordination*, as elicited by the several appropriate tests, both with eyes open and closed, was normal.

She had no adiadochokinesis, no dysmetria and no discoverable dyssynergia. She had no apraxia or dyspraxia. She wrote a fair hand with facility. There was no abnormality whatever in her speech, and her conversation was that of an intelligent and cheerful young woman.

The *deep reflexes* were normal, except the patellar and Achilles reflexes, which were considerably exaggerated. There was a transient ankle clonus on the right side.

Of the *superficial reflexes* the abdominals were absent, and there was a definite dorsal flexion of the right big toe, brought out by the Babinski, Oppenheim and Schäfer methods.

Her *muscular strength* was normal.

The *sensory examination* showed touch, localization, pain and temperature to be normal. There was a definite diminution of the vibratory sense in both lower extremities from the knees down. The sense of position was normal everywhere, except in the right big toe, where it was considerably impaired.

CRANIAL NERVES.—The olfactory and optic nerves were normal. Outside of a slight horizontal nystagmoid movement when the gaze was directed to the extreme right, the oculomotor, trochlear and abducent nerves showed no abnormality. The other cranial nerves were normal as well.

Her *mental status* showed nothing abnormal whatever.

Her *tegumentary, cardiovascular, respiratory* and *gastro-intestinal* apparatuses were apparently normal.

The blood Wassermann tests and the urinary examination made some time later, gave negative results.

On December 26 she was again attacked by paraplegia. Her legs became weak and heavy, until, in the course of a few hours, she could no longer walk or stand. This time the attack was accompanied by pain in the right side of the back, about the lower scapular region. The attack lasted only a few hours, and the next day she was as well as ever.

On January 14, 1921, the neurological examination showed the following: A somewhat greater spasticity of gait on the right side than that observed on previous occasions. A more definite Romberg. A more pronounced Babinski on the right and a rather doubtful one on the left side. A temporary ankle clonus on both sides, but greater on the left than on the right. A right and left tibialis sign of Strümpell. A slight diminution of pain and temperature sense over the left lower extremity and left lower abdomen.

Towards evening of the same day—January 14—her legs once more became weak and spastic. The trouble increased rather more gradually than on previous occasions and attained its height on the second or third day, when she was brought to the clinic in that condition. The neurological examination showed the following:

The patient could hardly walk without assistance on account of the spasticity of her lower limbs. She had a very pronounced Romberg. A pronounced dorsal flexion of both great toes was easily elicited by every known method. The suprapatellar, patellar and Achilles reflexes were excessively exaggerated on both sides. A permanent patellar and ankle clonus was easily induced on both sides. A tibio-adductor reflex, both patellar and periosteal, was present from left to right and from right to left. The pain and temperature hypaesthesia over the left lower limb and left lower abdomen was somewhat more evident than on January 14. The condition of the cranial nerves remained unchanged.

The patient was again examined on January 26. The symptoms of January 17 had entirely disappeared and so had the pain and temperature hypaesthesia of January 14. Instead two new symptoms made their appearance: a definite rotatory nystagmus when the gaze was directed to the left above a horizontal; and a flexion of the right thumb upon snapping the finger nails—Hoffmann's sign. The right Babinski and the absence of the abdominal reflexes remained unchanged. The originally barely perceptible spasticity of gait was now less perceptible than ever—the patient walked better than at any time since her appearance at the Vanderbilt Clinic.

DIFFERENTIAL DIAGNOSIS AND PROGNOSIS

Upon subtracting the transient and mutable symptoms observed in this patient for two months, we are left with a single residual sign of organic disease of the central nervous system—the sign of Babinski on the right side. Since, however, the intensity and duration of the other abnormal manifestations exhibited by this patient have varied with a degree of capriciousness which is inconsistent with our ideas of a permanent lesion in the central nervous system, we can by no means be certain that the sign of Babinski may not disappear any day. As to the absence of the abdominal reflexes, much as this symptom may assist diagnosis in the presence of other definite symptoms, it helps not at all in their absence. No one doubts Marburg's[1] conclusion that the absence of the abdominal reflexes is the most constant accompaniment of multiple sclerosis. But these reflexes are absent in a number of other conditions; and they are frequently difficult, and sometimes impossible, to elicit in a normal person with a large and flabby abdomen—and the abdomen of our patient is neither small nor firm.

The classical symptoms of multiple sclerosis—scanning speech, intention dyssynergia, definite nystagmus and temporal pallor of the optic discs—are wanting in this patient. Yet, while the sign of Babinski persists, we are inclined to diagnose the disease as multiple sclerosis rather than hysteria. But let that sign disappear, and the differential diagnosis between the two diseases in this case must be extremely difficult. What helps us out of the present dilemma is the vast number of parallel cases recorded by competent observers for the last thirty years. The conclusion at which one arrives on the strength of those observations is that the salient feature of multiple sclerosis is not so much this or that sign or symptom, but the extraordinary mutability and evanescence of all its abnormal manifestations, no matter how alarming those manifestations may be for the time being. One reads of Warrington's[2] cases, who are affected with blindness, supposedly due to optic atrophy, for months and years; who are now and then, in addition to their blindness, stricken down with various forms of paralyses and other alarming disturbances; and who most inconsistently suddenly appear in public as if nothing had ever happened to them. J. Dixon Mann[3] tells of patients who take walks of several miles between oft-recurring attacks of diplopia, strabismus, spastic paraplegia and fecal incontinence, and who, when last seen, appear to be quite well. When such patients have the organic lesions characteristic of multiple sclerosis, then we may reasonably assume that our patient, who displays a similar inconsistency of behavior, is similarly affected.

The transience and mutability of the patient's symptoms can to a large extent be explained in the light of a nerve lesion which, although it leaves the axis-cylinders denuded of their myelin sheaths, and perhaps a good deal distorted (A. Thomas[4]), does not entirely deprive them of the function of propagating nerve impulses. It stands to reason that such axis-cylinders are more vulnerable to the action of the poisons which now and then circulate in our blood for a time than are healthy and well-insulated nerve fibers. We can understand how a slight and temporary intoxication which will cause a headache or a "fit of blues" in a person with normally myelinated nerve fibers, may cause alarming, though transient, symptoms in a person whose nerve myelin is denuded in spots. The most prominent manifestation of our patient's disorder—the sign of Babinski —has been repeatedly brought about in perfectly healthy persons by temporary poisoning. The work of Link[5] with scopolamine injections, and that of Volkmann[6] with morphine-scopolamine injections, illustrate in a very convincing way the action of transient poisons upon the pathway which hold in abeyance the reflex dorsal flexion of the great toe. Whether the original cause of the sclerotic lesions is a specific infectious process or not, it is safe to assume that the subsequent manifestations of the disease are caused by the transient action of endogenous or exogenous poisons upon denuded, and therefore vulnerable, fibers in the central nervous system.

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DISCUSSION

DR. CORNWALL. The evanescence of the symptoms seems to me rather significant of the present tendency to feel that multiple sclerosis is an infection. In the absence of spinal fluid serology, considering the sudden onset and the severity, I should be inclined to think first of lues, despite the negative blood, unless it is ruled out by negative serology.

DR. GOODHART. This is just the sort of case that we are all likely to slip up on. If it is multiple sclerosis, it brings up some questions in regard to that disease which have not yet been fully met. The interesting thing is the peculiar evanescence of the symptoms. I have never seen a case of multiple sclerosis in which there was such a kaleidoscopic variation, almost under one's very eyes.

Of course, we all know that the patches of multiple sclerosis are not confined to any special part of the nervous system. This peculiar selection of one portion of the nervous system is very unusual. I have never seen it, but it proves that it can occur. It may be that this woman is not going on to the progressive stages of multiple sclerosis. It may be one of the cases in which there is chronic infection. Dr. Rosett's suggestion as to the character of the lesion, that with the lowering of resistance there was a likelihood of a poison seizing upon these points of least resistance, is a rational one.

DR. MCKENDREE. With reference to the sudden onset in this patient, I should like to cite a case of a young woman I saw recently. She had her acute onset of multiple sclerosis with an attack of mumps, and after two or three months the symptoms subsided completely, so far as we knew, for two or three years. Then, with the onset of an attack of bronchial asthma, the symptoms again manifested themselves. After three or four months of treatment, she became functionally well, although it is quite easy to demonstrate the presence of multiple sclerosis still. In the past few years I have seen perhaps three or four cases in which there has been marked sensory dissociation. I have seen the spinothalamic system involved in multiple sclerosis, and I have seen the posterior columns involved. Only recently a man was seen at the Neurological Institute who had definite signs of multiple sclerosis with hypaesthesia or diminution to thermal stimuli in the leg.

DR. WECHSLER. It occurs to me that in this case we might be dealing with a peculiar vascular disturbance or an anaphylactic reaction. Claudication or vasomotor disturbance of the spinal vessels may offer an explanation.

DR. OSNATO. It seems to me that the evanescence and mutability of the symptoms of multiple sclerosis are readily explained when one considers the pathology of the condition. It is not an axonal infection but an infection of the medullary sheaths, and the symptomatology is really secondary to the toxic absorption from these foci of infection. It is not until a long time afterward that the axis cylinder is destroyed. So that whatever produces the intensity of the infection, if it is an infection that causes the condition, and whatever determines an increase of the condition, such as the matter of the blood supply, would also make for an increase or decrease in the center of the pathology. If it were a primary degeneration of the axis cylinders, the condition would be stationary. The suspicion that multiple sclerosis might be of an infectious origin has never struck me so forcibly as in this case. It seems to me that until recently neurologists have not studied their cases sufficiently from a medical point of view. There is such a thing as overlooking the other branches of medicine, and a case in point is the study of the blood and spinal fluid chemistry. This is an important factor in differentiating tuberculous meningitis from other types. One can make an absolutely definite diagnosis just from the study of the chemistry of the cerebrospinal fluid.

And it seems to me that it would be worth while to study cases of multiple sclerosis along the same lines.

As to the matter of the nystagmus: Any time that we have nystagmus a careful Bárány investigation should be undertaken so that we can get some order out of the chaos in the interpretation of these findings. Careful study should be given to the plane in which the nystagmus is found. The mere mention of a nystagmus is worthless unless some detail is gone into.

Dr. Tilney. In connection with this interesting situation I would like to propound an important question: If this is a case of multiple sclerosis, what is multiple sclerosis? This year the entire problem of multiple sclerosis has been raised by the Commission of the Association for Research in Nervous and Mental Diseases, and it has fallen to me to formulate some special problems concerned in the experimental work to be done in this investigation. After giving the matter careful consideration, I have suggested the following questions:

1. Is there any *clinical constant* in multiple sclerosis? Is there anything in its course or its symptomatology which we can regard as a clinical constant? It seems to me at present that the clinical constant is the inconstancy of the disease. What has been considered the characteristic triad of symptoms—nystagmus, intention tremor and explosive or scanning speech, is by no means adhered to to-day.

2. Is there any *pathological constant* in multiple sclerosis? Personally, I think the histological picture is very protean, and that it is yet to be drawn in all its details. We have no final word in this respect as yet. I am certain that we have still to establish a pathological constant. Perhaps this is due to the fact that we do not get enough specimens and pathological material. In this connection, I am particularly impressed by the fact that the nervous system embryologically comes from the same tissue which gives rise to the skin, the ectoderm. We know with what a vast variety of changes and under what a great number of varying causes the skin is liable to show pathological alteration. The same thing may be true, to a large extent, in the nervous system.

3. Is there any *bacteriological constant* in multiple sclerosis? That was brought more into the foreground than any other problem in relation to this disease, largely because of the work which seems to indicate that the disease is transmissible from patients suffering with it. We know nothing definite as yet, but it looks to me as if the field in bacteriology opens the most promising door to the solution of the important problem of this disease.

4. Is there any *biochemical constant* in multiple sclerosis? This includes the study of the spinal fluid and brings up the question of protein sensitization.

So in the question, What is multiple sclerosis? I think we must admit that we are all still largely in terra incognita—clinically, pathologically, and from the standpoint of etiology.

Dr. Casamajor. Certainly this case we saw to-day cannot be pathologically like the ordinary case of multiple sclerosis. When I first saw this patient the impression I got was that she had multiple sclerosis with a rather rapid onset. But to-day those symptoms have all disappeared. Of course there are fluctuations in multiple sclerosis, but we cannot explain her symptoms by a sclerosis—by an increase in neuroglia, for neuroglia fibers do not come and go like the tides. What has happened here is not multiple sclerosis, but probably a multiple toxicosis. I think that Dr. Wechsler came nearest to the solution when he called it an anaphylactic reaction. Dr. Tilney spoke of the connection between the skin and the nervous system. Possibly we shall be able to separate this condition which we call multiple sclerosis into a group of things, as in the skin conditions. If we could conceive of the nervous system reacting in an anaphylactic way to toxic substances as the skin reacts to urticaria, then we could gain some conception of the state of affairs existent in this patient's central nervous system.

Let us not try to find a pathology that will fit the whole thing. Let us first try to find out what multiple sclerosis is.

EPIDEMIC ENCEPHALITIS OR CEREBRAL SYPHILIS?*

A Case Presenting an Interesting Differential Diagnosis

By CHARLES A. McKENDREE, M.D.

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I am presenting the following case because it suggests a particularly interesting differential interpretation. At first examination I was reasonably sure of the diagnosis, but observation of the patient over a period of six months has raised certain doubts as to the correctness of the first impression. The history and development of the patient's symptoms combined with certain atypical findings are of sufficient interest to warrant our discussion of the case, especially from the standpoints of etiology and treatment.

Patient, Mr. W., American, aged fifty-four, married; occupation, actor. V. C. No. 29745.

Complaint.—Inertia. Excitability, such as trembling when he is planning to go anywhere or do some special thing. No ambition. No desire to do anything or go anywhere. Twitches during sleep. Spells of confusion, during which he cannot quite collect himself, cannot recollect the date, the day of the week, etc. Nauseated sometimes during these spells, but never vomits. At times he may ask "Where were we yesterday?" If told, he may reply that "it seems as though it were a year ago."

Duration, thirteen months; onset, acute; progress, about stationary in past ten months, but attacks of confusion are becoming more frequent of late.

Family History.—Mother died recently at eighty-seven, as the result of a stroke. Father died twenty years ago from pneumonia. One sister alive and well. Nothing similar in collateral branches. Wife has had one miscarriage (her blood Wassermann is said to be negative).

Past History.—No serious illness of any kind. Scarlet fever as a child. Tonsillectomy thirty years ago. Has always been considered a healthy man. Thirty-eight years ago the patient had an initial lesion. Was treated from the beginning for three years with mixed treatment. For periods since then he has resumed treatment for a few months at a time. Formerly he had frequent attacks of pharyngitis, but no serious trouble since his tonsils were removed. No accidents, no head injuries, no operations except tonsillectomy. No motor disturbances, no convulsions, no diplopia, no speech disturbances, no visual disturbances, no vertigo, no dysesthesia, no pains, except infrequent headaches. No vomiting up to present illness. Alcohol always in moderation. Has smoked in moderation. Tea and coffee not in excess.

Present Illness.—Was perfectly well up to thirteen months ago. He then complained of headache for about two days, rather severe at first. The second day he vomited several times, and had diarrhea. There were no nasopharyngeal symptoms. On the second and third days his wife observed trembling of the patient's lips, and spells of weakness and drowsiness. He had a low fever varying between 99° and 100° for about a week. He slept well every night, but also during the day. He was allowed to be up and dressed, but he would be inclined to lie down and doze for short periods frequently during the day. During the periods when his lips would tremble, his eyes would fill with tears, although he did not have any emotional upset. The drowsiness during the day continued for three months in marked degree, but since that time, in spite of an inclination to sleep, he cannot always get to sleep when he lies down during the day. For the past ten months he has been troubled with insomnia. He lies awake at times until 6 A. M. As a rule he gets to sleep about 3 A.M. (The patient usually does not retire before 1:30 because of his work, which necessitates his keeping late hours.)

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*Presented at a Conference of the Neurological Staff, Vanderbilt Clinic, February 28, 1921.

No diplopia, and no facial weakness have ever been observed. In the first week of his illness he was somewhat delirious, but not at any time since. His wife says that there have been no delusions, no hallucinations, or other abnormal mental phases, but in the past thirteen months his memory has been affected for recent events, though not for past. He recalls events of twenty years ago perfectly. He has difficulty in concentration, as for example, in writing letters. He reads the newspapers, but says he does not retain well what he reads. He can memorize, however. Some months ago he memorized several poems and small parts, which he has retained perfectly up to the present. (Patient recited one for me without noteworthy error.)

He is depressed most of the time, disinclined to talk or mingle with others. He recognizes the change that has taken place, and is quite discouraged.

For the first nine months he was unable to do any work, but recently he has been able to do "extra work" on the stage. This does not entail any speaking parts, but appearing in crowd scenes, etc. Very infrequently the patient experiences a sensation of well-being which soon passes off. There has been no expansiveness and no exaltation of thought or of manner.

The symptoms enumerated above have been present off and on since the acute onset, and his wife is very definite in asserting that up to that time the patient was absolutely well.

Appetite has been poor recently. Bowels quite regular. No loss in weight. No resppiratory symptoms. No urinary difficulty of any kind. No sexual desire since his illness. At my suggestion the patient has been receiving intravenous salvarsan treatments in this department since last September.

EXAMINATION

VOLUNTARY MOTOR SYSTEM.—There are no abnormal attitudes or deformities. The gait is normal in every respect. Equilibratory and non-equilibratory tests for upper and lower extremities present no deviation from the normal. No dysmetria.

Skilled acts.—Speech is normal, both spontaneous, and in test phrases.

Abnormal Involuntary Movements. — Careful observation reveals no tremors except very slight movement in the tongue. No other abnormal movements present.

Reflexes

DEEP	*Right*	*Left*
Tricipital	2	2
Bicipital	1	1
Radial	1	1
Patellar	1 plus	1
Achilles	1 plus	1
SUPERFICIAL		
Supra-umbilical	0	0
Suprapubic	2	2
Upper lateral abdominal	1	1
Lower lateral abdominal	2	2
Cremasteric	1	1
Plantar flexion	0	0
Babinski	0	0

Muscle Strength.—There is no detectable weakness in any of the muscle groups.

Muscle Status.—There is no atrophy, and no hypertrophy. Tone is quite normal. Consistency normal.

Abnormal Associated Movements.—None noted.

GENERAL SENSORY.— Touch, pain and temperature sensibility is normal throughout. Vibratory and muscle-tendon sense unaffected in upper and lower extremities. Stereognosis, normal.

CRANIAL NERVES. *Optic, Oculomotor, Trochlear and Optic Apparatus.*—Fundi normal. Pupils equal, practically round, normal in size, central. They react to light rather slowly on each side, and the excursion is not as wide as commonly seen. To convergence there is prompt and normal reaction. It is questionable whether the disparity between the light and convergence reactions is sufficient to call the pupils of the Argyll-Robertson type. There is no strabismus, no ptosis, no exophthalmus or enophthalmus. Slight exophoria of left eye in convergence tests. No nystagmus. Palpebral fissures equal.

Acoustic and Ear.—Not tested. Hearing apparently normal.

Trigeminus and Mouth.— Motor status normal. Teeth show many fillings. Considerable leukoplakia.

Facial and Face.—Motor status normal. Articulation of labials normal. Associated movements of face, eyes and head normal. Vasomotor and secretory status normal.

Glossopharyngeus, and Vagus, Pharynx and Larynx.—Uvula moves normally. Phonation normal. Swallowing normal.

Spinal Accessory.—No deviation from the normal.

Hypoglossus and Tongue.—Tongue protrudes in the midline; is placed in each cheek normally. Trophic status normal.

MENTAL STATUS.—The patient is intelligent, cooperative, and presents no emotional instability. He is quite conscious of his diminished memory for recent events. He recites poetry learned several months ago, without many errors. There is no obtainable evidence of hallucinations, delusions, or other psychotic manifestations.

SYSTEMIC EXAMINATION.—*Tegumentary status* normal.

Glandular Status.—Thyroid normal. Tonsils present but show no noteworthy abnormality. No cervical or inguinal adenopathy. An epitrochlear gland can be felt on the left but not on the right.

Cardiovascular.— Pulse normal in rate and rhythm. Systolic pressure 148 mm. Heart normal.

Gastro-intestinal.—Nothing abnormal detected.

Genito-urinary.—No urinary disturbance. Sexual desire absent for past thirteen months.

LABORATORY TESTS.—*Blood Serum*: Wassermann four plus positive.

Spinal Fluid: Globulin, two plus. Wassermann negative to 2.0.

SUMMARY OF IMPORTANT SYMPTOMS AND FINDINGS

1. Acute onset, thirteen months ago with headache, low fever, vomiting, diarrhea, drowsiness and nervous symptoms.
2. The persistence of drowsiness, with insomnia of varying intensity.
3. Decided change in personality; poor memory for recent events; attacks of confusion; excitability; apathy.
4. Questionable inequality in deep reflexes of lower extremities; questionable Argyll-Robertson pupils; loss of sexual desire.
5. Positive Wassermann reaction of blood serum. Negative spinal fluid.

DISCUSSION

The acute onset of our patient's illness at a time when epidemic encephalitis was at its height, together with symptoms quite characteristic of certain types of the disease, naturally suggests one diagnosis. Certainly if we did not have a definite history of luetic infection with positive blood findings, we would be quite satisfied with such an explanation. For although the mental picture presented here has not been described to my knowledge, all of us have been brought face to face with similar atypical mental states following encephalitis. We are all familiar with the various psychotic and nervous phases of the onset, but sufficient time has not yet elapsed for establishing our information as to the mental sequelae of extensive involvement of the central nervous system in certain cases. We are familiar with the Argyll-Robertson pupil and other organic sequelae of this disease. Hence when we are confronted with such a picture, combined with a positive Wassermann, the problem should enlist our special interest.

General paresis, of course, must be seriously considered in this case. Progressive failure of the mental powers, failure to apply one's attention continuously, lapses of memory, irritability, restlessness, depression or excitement, may characterize the early stage. But in the majority of cases we usually find more outstanding symptoms after the lapse of a year. We have seen that there is no expansiveness in this patient, no real deterioration, in fact, none of the classical signs and symptoms of advanced paresis. Personally I do not believe that the treatment to which he has been submitted could have prevented the exhibition of further signs and symptoms, if the underlying process were that of paresis. The results of the examination made this week are identical with those of six months ago. Unfortunately only one spinal fluid examination has been obtained, and in the fluid previously submitted no cell count or Lange test was made. But the Wassermann of the fluid was negative. This fact alone is of no value, but I believe that when the clinical features are considered with it we can be quite sure that we are not dealing with paresis as a clinical entity.

If we disregard the acute onset as being due possibly to some coincidental infection or toxemia, we still have to account for the drowsiness which persisted for so long a time. The persistent insomnia during the past ten months, which developed acutely, after three months of normal sleeping and many periods of sleep during the day, is not easily explained. I have been unable to satisfy myself entirely as to the existence of an epidemic type, but it is difficult to explain some of the symptoms by any other interpretation.

Vascular syphilis of the brain seems to be the most acceptable diagnosis in this case. The patient is having more frequent "spells of confusion" although his other symptoms remain practically stationary. To my mind, these suggest vascular disturbance of a temporary nature. The change in personality is undoubtedly due to organic changes, the result of vascular disease. The positive blood and negative fluid findings would bear us out in this interpretation. Personally, I have never encountered vascular syphilis manifesting itself in such an abrupt fashion, with fever and persistent drowsiness. I should be very glad of the opinion of the conference upon this point. If this patient had an encephalitis, as would seem probable, it is interesting to note the development of cerebral syphilitic lesions upon the basis of an acute infection.

DISCUSSION

Dr. Cornwall. I have been treating this man for six months and until to-day, when the case was presented, I had not connected the acute attack that he had some time ago with his present condition. I had labeled him a case of vascular syphilis of the cerebral type and believed that that was sufficient to account for his lapses of memory as well as some of the other symptoms that Dr. McKendree has touched upon or that his wife has explained in detail. I did not think that he was a case of paresis. Because of the frequency with which we see cases of neurosyphilis that have certain mental disturbances, slight depression, anxiety, moderate tremors of the tongue, with inability to concentrate, and the tendency that there is to call them all paresis, I have been calling them parenchymatous neurosyphilis, believing that that term covers the pathology.

I do not know how we are going to distinguish the diagnosis of cerebral vascular syphilis from encephalitis when the two diseases occur at the same time, unless cases of encephalitis reported by the early authors on this subject might be utilized by us in a study of the sugar content of the cerebrospinal fluid. In going over some of the early cases I find that some were reported as encephalitis, having in the absence of diabetes an increase of sugar in the cerebrospinal fluid. I think it would be interesting to study cases in that light now.

Dr. McCumber. In the absence of unequal and irregular pupils, and the fact that there are no marked fibrillary tremors, I do not think that we can make a diagnosis of paresis. I think it is more likely a vascular syphilis.

Dr. Sands. The first picture that came to me was that of a cerebral arteriosclerosis of the Binswanger type. However, the patient has a definite history of an initial lesion, and a four plus Wassermann. Furthermore the fact that he has no changes in the retinal vessels might dispel the idea that this was a case of the Binswanger cerebral arteriosclerosis. The next thing was a diagnosis of encephalitis. Whether he has that condition or merely a syphilitic endarteritis is important. I feel that he has more than an ordinary cerebral vascular syphilis. Cerebral vascular syphilis invariably comes on a little earlier in life; it is a mesodermal syphilis. The fact that he does not show more of general paralysis does not mean much. I feel that he presents the condition which Pearce Bailey described some time ago as the pre-paretic period of general paralysis. The fact that he has had negative Wassermann does not mean much because one examination is not sufficient. I would like to have three Wassermanns of the spinal fluid made by three different persons. We have now a case in Bellevue brought in to prove that he has nothing more than a traumatic neurosis. The usual tests except spinal puncture had been made. At one time a two plus blood Wassermann had been obtained in our wards. The ordinary routine lumbar puncture was done and the result was a four plus Wassermann. The blood gave a one plus reaction.

This case to my mind, while it is little more than ordinary cerebral vascular lues, belongs, I think, to the pre-paretic group of syphilis.

Dr. Polon. The patient's incapacity to work, his changed personality and absence of localizing signs, most likely speak for a parenchymatous change in the brain. The prominent symptoms are sluggish pupillary reaction to light, positive Wassermann and changed personality, and all that follows what appears to have been an attack of lethargic encephalitis. Time and again observers have reported the initiation of a general paralysis following this disease. I would therefore conceive this case to be what Dr. Sands has termed a pre-paretic state initiated by the attack of encephalitis the patient had.

Dr. Wechsler. This man undoubtedly had an encephalitis. Frequently we see psychic changes following encephalitis, so why should we blame the luetic process for these changes? There is no other symptom in favor of paresis. His confusional state is not a very pronounced one. I think this is a distinct encephalitic process which has left in its wake certain slight mental symptoms, and I should think that the prognosis would be good. I should leave out further specific treatment. For one thing, I do not think it will help him and for another it will confirm him in the thought that he has lues.

Dr. Rossett. I am inclined to agree with Dr. Wechsler. At present I do not see why we should say it is pre-paresis, although he may have syphilis and may have paretic symptoms in time.

Dr. Lehman. I agree with Dr. Wechsler. I do not think there are enough physical findings or mental changes to label this man a paretic.

Dr. Tilney. We know this man has syphilis. His cerebrospinal fluid may not be positive but his blood is, and that is something which we must not overlook in our treatment. No one would presume to say that there is not some involvement of his central nervous system by the syphilitic process. Whether he has had encephalitis or not, he should have antisyphilitic treatment.

Dr. Casamajor. I think the important thing to bear in mind is that this man undoubtedly has syphilis. As to the suggestion of having Wassermann tests made at several laboratories, personally, I am against this. If you have enough examinations made you can get a positive Wassermann from some place. But that often means that an individual is trying to prove something. When you have a report from one good laboratory, that is enough. If you increase the number of laboratories you increase the chances of error.

The patient has syphilis; he has a changed mental state, a change of personality. It is not a paretic type of deterioration. It is much more an arteriosclerotic type. The mental changes he shows are not the changes that have been described in epidemic encephalitis, nor are these periods of confusion at all reminiscent of a neurotic state. I think this man has an organic disease of the brain with intermittent variations which make it look as though it is due to a vascular lesion. I am not at all enthusiastic about calling these conditions preparetic stages. That I think is too fine a point to make clinically. Nor do I think that we can speak of parenchymatous syphilis as a thing apart from the rest.

General paresis means a clinical entity, a definite prognosis, and leaving out the question of legal medicine altogether, I do not see how we can get away from the term general paresis. I am sorry we did not get a gold curve in this case, it might have helped us. We might have gotten that typical curve which is usually seen in general paresis.

I think the question as to whether the man had syphilis is a question much more nearly settled than if the man had encephalitis. The arterial changes may well have given us all the things that Dr. McKendree has spoken of. Certainly we see cases of cerebral syphilis which are diagnosed as epidemic encephalitis. I have seen three cases of fractured skull that were diagnosed as epidemic encephalitis. I would be much more inclined to doubt that he had epidemic encephalitis than that he had syphilis.

Dr. McKendree. I would like to mention one little incident which is against this being a case of psychoneurosis. During one of his periods of confusion he asked his wife: "Where are our dogs?" She replied, "We never had any dogs," to which he answered, "Oh, I see." That one phrase, I think, takes him out of the psychoneurotic class.

EPIDEMIC ENCEPHALITIS OF A PARALYSIS AGITANS TYPE WITH A RECRUDESCENCE ONE YEAR LATER

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At one of the Neurological Conferences held in December, 1920, at the Vanderbilt Clinic, a case was shown of epidemic encephalitis with a progressive paralysis agitans syndrome. The case presented to the conference to-day, although showing symptoms of paralysis agitans, gives signs other than those of the extra-pyramidal system. Also of especial interest is an exacerbation of the symptoms following one year's apparent improvement in health.

J. G., patient in third male medical ward of the Post-Graduate Hospital, age nine years, white, American, of Italian descent, schoolboy.

HISTORY.—The present illness began about one year ago, January, 1920, with quite sudden onset. The symptom first noticed was fever accompanied with a malaise state. At the time of onset, there was projectile vomiting, which continued irregularly for several days; this was not preceded or accompanied by nausea. The patient was constipated. There was difficulty in voiding urine, which disturbance lasted for several days. A urinalysis made at the time showed a hematuria. In a few days the febrile condition was accompanied with delirium and a complaint of intense pain in the sub-occipital and post-cervical regions, also pains in the left arm and leg.

The symptoms gradually subsided and at the end of three weeks the patient was out of bed. When he began to walk, an unsteadiness of gait and a tremor of the left hand and foot were noticed. Diplopia was also complained of at this time. The parents observed an emotional change. The boy was impatient and irritable, had fears and cried often without sufficient cause.

During convalescence he has progressed quite well, the body strength improving. He has had a good appetite and has slept well, but he has been more emotionally unstable than before the illness and the tremor of the hand and foot has never entirely disappeared. Because of this condition, he has not returned to school during the past year.

On January 19, 1921, one year after the initial symptoms of his illness, the patient was quite suddenly taken ill with a fever and vomited at infrequent intervals; the vomiting was of the projectile type, without nausea. There was no constipation. The bladder became distended with urine and there was difficulty in voiding. The tremor of the hand increased and the emotional state became more unstable.

On January 26, 1921, the patient was admitted to the Post-Graduate Hospital on Dr. Tilney's service as an ambulatory case, with the following complaints: tremor of the left hand and foot, slight unsteadiness in gait, post-occipital headache and unstable emotional state.

Injuries and Diseases.— Tonsillectomy performed in 1915. Measles at four years of age.

Habits and general physiological status.—Drinks a cup of tea daily. Kept awake during the day, but sleeps poorly during the night; is usually wide awake until 3 or 4 A.M. Appetite is good, bowels have been regular, nocturia two or three times a night.

Family History.—Father, age forty years, alive and well; mother, age thirty-seven years, alive and well, four brothers and two sisters alive and well.

VOLUNTARY MOTOR SYSTEM.—*Abnormal attitudes and deformities:* Stands erect. The left leg and thigh are slightly smaller in circumference than the right.

Gait: He walks slowly with a very slight tendency to drag the left leg. The head is held slightly stiff with a slight anterior flexion; there is an impaired associated movement of the left arm. He stands on toes well, but is unsteady when he stands on heels.

Coordination: Patient is right-handed. Equilibratory: He stands normally with feet together, eyes open. Romberg is absent. Stands normally on the right foot, but has slight unsteadiness when standing on the left. Non-equilibratory: Finger-to-nose, finger-to-finger, each finger-to-thumb tests are performed normally on the right side. On the left there is a possible slight ataxia and this is accompanied with a slight tremor. Finger-to-thumb—this is done more slowly on the left. Pointing and pass-pointing are performed normally on the right and left. Heel-to-knee and along-shin tests are performed normally on the right, fairly well, but more clumsily on the left. Adiadochokinesis: Right is absent, left is present. Dysmetria: Absent on right and left.

Skilled Acts.—No apraxia or aphasia. Speech is of slow rate and in a monotonous tone.

Abnormal Involuntary Movements.—There is a slow, coarse, regular tremor, of equal force and amplitude, of the left hand and foot seen at rest. It is of greater amplitude in the hand than in the foot, is decreased by volitional effort and increased by emotion. No tremor is seen in the right upper and lower extremities.

Reflexes:

DEEP	*Left*	*Right*
Jaw	Active	Active
Pectoral	Active	Active
Biceps	Active	Active
Triceps	Active	Active
Radial	Active	Active
Ulnar	Slight	Slight
Wrist	Active	Active
Suprapatellar	Active	Active
Patellar	Very active	Active
Patello-adductor, R to L	Absent (L to R)	Absent
Tibio-adductor (periosteal)	Active	Slight
Tibio-adductor, R to L	Absent (L to R)	Absent
Cuboido-digital (dors. flex. toes)	Active	Active
Achilles	Very active	Active
Ankle clonus	Transient	Absent

SUPERFICIAL	*Left*	*Right*
Ciliospinal	Present	Present
Supra-umbilical	Active	Active
Suprapubic	Active	Active
Upper lateral abdominal	Active	Active
Lower lateral abdominal	Active	Active
Cremasteric	Active	Active
Plantar flexion	Absent	Active
Babinski (toe)	Present	Absent
Babinski (fanning)	Present	Absent
Chaddock	Present	Absent
Gordon	Absent	Absent
Oppenheim	Absent	Absent
Schäfer	Absent	Absent

Muscle Strength.—He is able to rise from recumbent position to sitting and from sitting position to standing without difficulty. Opposed movements of the left upper and lower extremities show a degree of strength slightly less than normal.

Muscle Status.—There is a slight atrophy of left thigh and leg muscles and a lessened consistency of the left thigh and leg. The tone as tested by extensibility of the joints shows a cogwheel movement of the left elbow.

Abnormal Associated Movements.— A doubtful Hoffmann sign and the thumb sign of Klippel-Weil are present on the left. The sign of Grasset-Bychowski is absent.

GENERAL SENSORY.—Touch and touch acuity, pain (superficial and deep), temperature (cool and warm), vibratory and muscle-tendon sense and stereognosis are normal.

CRANIAL NERVES.— *Olfactory:* Smell (subjective) normal.

Optic: Oculomotor, Trochlear, Abducens Nerves and Optic Apparatus. Vision, left and right, acuity and fields are normal. Fundi are normal. Pupils, left is less than right (L. 3 mm., R. 3.5 mm.) in diameter. Shape, both pupils are somewhat oval in shape. Position is normal. No hippus present. The reaction to light, direct, left and right, is prompt and of good amplitude; consensual, both left to right and right to left, is normal, and there is no Argyll-Robertson pupil. Position of the eyes is normal and there is no diplopia at present. There is no limitation of voluntary movement. There is a nystagmoid movement, lateral in direction and of small amplitude to the right; this is also present slightly to the left. Palpebral fissures are equal. No abnormalities of iris, lens, media, cornea or conjunctiva.

Acoustic nerve and ear: (a) Cochlear, hearing, subjective and objective is normal; (b) Vestibular, caloric, rotatory and Hitzig tests not made.

Trigeminus Nerve and Mouth: Masseter of normal strength. Corneal reflex, left and right, is normal. Teeth are irregular in position.

Facial Nerve and Face: Taste, subjective, is normal. The left lower facial muscles seem slightly flattened with a slightly delayed movement of left lower facial group on voluntary motion. No change in the vasomotor and secretory status.

Glossopharyngeus and Vagus Nerves: Taste is normal, position of uvula central and the movements of the palate during phonation are normal. The voice is of a monotonous tone. There is no difficulty in swallowing.

Spinal Acessory Nerve is normal.

Hypoglossus Nerve and Tongue: The tongue is protruded in the midline, and the movements in all directions are normal.

MENTAL STATUS.—The impression as to intelligence is that he is normal, but there is a manifest emotional instability.

SYSTEMIC.—The skin is smooth and moist and he perspires freely over body. Tonsils are enlarged and a few lymph glands are palpable in the cervical region. Cardiovascular: a systolic murmur is heard at the apex and base.

Respiratory, gastro-intestinal and genito-urinary systems were negative at the time of examination.

LABORATORY EXAMINATIONS

January 27, 1921; urinalysis showed the urine cloudy, sp. gr. 1.020, alubmin and sugar negative, microscopical examination negative. February 2, 1921; color amber, reaction alkaline, sp. gr. 1.022, albumin negative, sugar present, 4%. Microscopical examination negative. January 28, 1921; blood Wassermann negative; spinal fluid Wassermann negative; colloidal gold normal; the spinal fluid shows 1 cell per c.c. and no increase of globulin.

SUMMARY AND CONCLUSION

A boy in good health was suddenly taken ill with fever, malaise and projectile vomiting. This was quickly followed by delirium, a distended bladder, with a loss of expulsive power and a hematuria. There was suboccipital pain, also pains in the left arm and leg. The symptoms gradually subsided and the patient was out of bed at the end of three weeks. He then first complained of diplopia and the parents noticed a constant tremor of the left hand and foot and an unsteadiness in gait. He was emotional. The boy's health improved, he was out of doors and played with other children, but there was a residual non-intention tremor and the associated movements of the left hand and foot were impaired. He has remained very emotional. At the end of one year, he developed a low-grade fever with projectile vomiting, distended bladder and increased tremor of the left hand and foot. There were also rather intense emotional reactions and headaches. The neurological examination showed signs of a paralysis agitans syndrome and some pyramidal tract signs. With two weeks' rest the tremor has slightly diminished and the patient has gained two pounds in weight. The initial symptoms one year ago are interpreted as clinical manifestations of an acute inflammatory process with a moderately severe toxemia involving the cerebrum and basal ganglia. This process, or the pathological changes following, have never entirely subsided and one year later there is an exacerbation of the symptoms with gastro-intestinal and bladder disturbances and emotional reactions as in the primary attack. Among the large number of cases of epidemic encephalitis there are many which apparently entirely recover, but the study of this case leads one to question how long following convalescence there may be a relapse or recrudescence of symptoms in this disease.

DISCUSSION

Dr. Tilney. I do not think there is any question as to the diagnosis in this case. The point that interests me most is the long period between the initial attack and the second outburst of the symptoms. It seems to me there is a possibility, already mentioned a number of times, of the organism remaining somewhere in the body in a state of latency. These recrudescent outbursts of the symptoms resemble what we are familiar with in syphilis of the nervous system, and it makes one think that the particular pathogenic agent must be held in a state of rest somewhere. We have already discussed the two most important possibilities that have been raised up to this time: first, that the organism is held in the perivascular lymph spaces and then, under some stress, becomes active again; second, that these patients may be carriers of the disease in the mucous membrane of the upper respiratory passages for a much longer period than we think.

I believe perhaps one of the first procedures that should be followed in all cases of epidemic encephalitis, after the acute symptoms have subsided, is to make a washing of the nasopharynx and perform a test to find out whether any viable organisms of the disease are lodged in the mucous membrane. I would certainly advise this test in the light of the many cases we have seen in which the patient has been pronounced recovered and yet the symptoms return sometimes with greater intensity than they had in their initial attack.

That is the practical side of this particular discussion. Now as to the localization of the lesion. It seems to me we are dealing here with exactly what Dr. Blakeslee has considered it, namely, an involvement of the striatal system extending, as it well may, into the projection system of the pyramidal tracts and thus accounting for the coexistence of pyramidal and extrapyramidal symptoms. The emotional instability probably indicates an extension of the more severe lesions into the region of the optic thalamus. So we have another one of those instances in which we see combined in a single patient the possibilities of involvement near the base of the brain, including the optic thalamus which controls the emotional status, the pyramidal tract, and the striatum, producing the symptoms in this case of paralysis agitans type.

Dr. Osnato. There is one thing about this case that might be mentioned: This second attack was ushered in by an acute coryza. That emphasizes the possibility of an actual reinfection.

Another point: In his attack of more than a year ago he had retention of urine, and now with the second attack the most prominent symptom at first was the trouble with micturition. This is interesting from a physiological standpoint. There has always been some discussion as to whether there may not be a cortical center of micturition. This boy showed in the beginning delirium and emotional changes, purely cortical and pyramidal manifestations. The only form of paralysis that he shows now is an upper motor neurone type. Many cases of epidemic encephalitis show just that thing.

The emotional changes in encephalitis have been likened to those seen in dementia precox more than in anything else. This boy, for instance, will cry with apparent great fear one minute, and the next minute will smile and play with everybody in the ward.

[It was suggested that a colloidal test be made in this case because meningitis had not been ruled out and the illness had started with a hematuria. The results of a colloidal gold test might indicate whether the condition were meningitis or encephalitis.]

Dr. Blakeslee. I think there is no doubt about this being encephalitis. I saw a great many cases of meningitis when I was in the army. But this one differs from them. As to the colloidal gold test as a means of differentiation between meningitis and encephalitis, I do not agree with that belief at all.

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THE ANATOMICAL AND CLINICAL SYNDROMES OF THE CORPUS STRIATUM*

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INTRODUCTION

The functions of few structures forming part of the brain have remained so veiled in mystery as that of the corpus striatum. Up to recent years the question was still being asked, "What is the influence of these central grey masses upon the innervation of the body?" Some authors denied that they played any appreciable part in the activity of the nervous system. It was useless to search French or foreign textbooks of neurology for a chapter dealing with the pathological symptomatology produced by lesions of the caudate and lenticular nuclei. It was therefore not at all unexpected to find a neurologist like Edinger[1] astonished at the limitation of our knowledge of the functions of the corpus striatum. About ten years ago the Frankfurt neurologist expressed himself as follows: "There is scarcely anything which shows more clearly how faulty our powers of observation have been than the fact that we know nothing of the functions

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of the corpus striatum and that we know nothing of the symptoms resulting from its destruction or irritation."

This mystery must have especially interested an anatomist like Edinger since he had made a special study of the corpus striatum in the vertebrate series. For a long time he had recognized its importance and constancy in the general plan of the nervous system. It is difficult to understand how an organ occurring in fish and reptiles, forming the major part of the cerebral hemispheres in birds, and myelinizing so precociously in the human fetus that the fibers of the ansa lenticularis are surrounded by a myelin sheath as early as the fourth month, could fail to have any appreciable function. How can we admit that the destruction which so often occurs in the human corpus striatum is not manifested by identifiable symptoms? Of course, one could reply that in man the striate body constitutes a system undergoing regression and that its function, mysterious in lower animals, tends to become more and more effaced by the preponderance of higher centers. That this is a most specious argument is established by plain facts. C. and O. Vogt[2] pointed out that the striate body, from the structural point of view, has none of the characteristics of a rudimentary organ. We cannot establish, in the striate body of man, any modification of structure as compared to that of the cercopithecus,* for example. Despite the most careful researches, no trace of regression can be found. From the anatomical point of view alone everything points to the fact that the striate body must possess important functions. This makes it reasonable to believe that our estimate of the importance of its physiology should be more extensive than has been in the past.

The scarcity of physiological and clinical data bearing on the striate body does not mean that many persons have not concerned themselves for some time past in unveiling the mysteries of its functions. The results of these researches have been very uncertain and often very contradictory. To make even a succinct analysis would take us too far and would, to any who read it through, be rather sterile and fantastic. After reading the greater part of these works one gets the impression of sterile efforts or uncertain opinions.

In reality, light has been shed on the part played by the striate bodies since the first researches of Madame Cecile Vogt by Kinnier Wilson,[3] Ramsay Hunt,[4] Cecile and Oskar Vogt[5] and Zingerle[6]. Thanks to the work of these authors we possess a clue which permits us to find our way through the labyrinth of anatomico-clinical facts accumulated during many years, and to have a more certain interpretation of the functions of the basal nuclei of the brain. It is worth remarking that we owe our most certain knowledge of the functions of the striate body almost exclusively to the anatomico-clinical method. Our knowledge of its physiology has not made such rapid strides. In attempting to state precisely the physiological rôle of the lenticular and caudate nuclei, we must rely on the facts which have been given us by semiological analysis controlled by anatomical and histopathological examinations.

Before getting to the heart of our subject it seems necessary to sketch rapidly the structure of the organs whose functions we wish to study. It is necessary to call attention to the fact that the architectural and histological organization of the nervous system is but the reflection of the life that animates it and of the functions which unfold themselves therein. Morphology has very often pointed out the way to physiological and clinical investigations. If there were any doubt of it, the history of the evolution of our ideas concerning the functions of the striate body would serve as a proof in favor of this opinion which we have expressed.

A short summing up of our knowledge of the structure and connections of the striate body in man seems worthy of our attention. We are indebted to the very recent researches of C. and O. Vogt, Bielschowsky,[7] Malone[8] and Ramsay Hunt[9] for data indispensable to the comprehension of certain anatomico-clinical syndromes which we have in view. This will be our excuse for penetrating the arid soil of anatomy and exploring that which, even yesterday, was considered a part of the vast *terra incognita* of the brain.

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* Cercopithecus: A species of monkey. The Ethiopian representative of the Asiatic macaques. *Encyclopedia Britannica*, XII, 670.

II. HISTOLOGY AND CONNECTIONS OF THE CORPUS STRIATUM

A. Introduction.—The corpus striatum is composed of two elongated masses at the base of the brain. These masses are two nuclei, distinct in appearance; the caudate nucleus, intra-ventricular, and the lenticular nucleus, extra-ventricular. In reality this morphological distinction does not correspond in any way to the real separation of these nuclei.

In some cross sections of the brain the lenticular and caudate nuclei appear very distinct, isolated as they are by the anterior limb of the internal capsule. Other sections taken lower down show fusion of the external segment of the lenticular nucleus with the caudate nucleus. This external segment of the lenticular nucleus, the putamen, belongs, from its appearance, to the caudate nucleus and may be distinguished microscopically from the two internal segments of the lenticular nucleus, which together form the *globus pallidus*. There is a great deal more than a resemblance. The recent researches of Vogt, Bielschowsky, Ramsay Hunt, Malone and others, have shown the fundamental fact that the putamen has the same histological structure as the caudate nucleus. The putamen and caudate nucleus are formed by an interlacing of the fine nerve fibers in the interstices of which are found two varieties of nerve cells. The first variety consists of the cells called type I of Golgi, with long axis-cylinder, and abundant protoplasm, through which are scattered the chromatophile bodies of Nissl, highly colored and filled with lipochrome (Bielschowsky); the second variety is made up of nerve elements called type II of Golgi, with short axis-cylinders, many richly ramified dendrites, whose small amount of protoplasm contains no chromatophile bodies. Bielschowsky has pointed out that around the cells of type I neuroglial corpuscles very often accumulate showing the fragility of these elements. The same author showed, moreover, that very often the vessels of the *putamen* are surrounded by a calcareous carapace unaccompanied by any appreciable symptoms attributable to this condition.

Quite the opposite, the *globus pallidus* is made up exclusively of a single variety of cells, those of Golgi type I, whose characteristics are mentioned above. The axis-cylinders which are derived from them are extremely long and cannot be followed for more than a small part of their length. Large numbers of endings of *unmyelinated* nerve fibers coil around the surface of these cells (Bielschowsky). From this description it follows that the *caudate nucleus* and the *putamen* belong in the group of *allomorphic* nuclei in the sense of Kohnstamm and the *globus pallidus* in the group of *isomorphic* nuclei.

Though the histology of the lenticular and caudate nuclei can be schematized in this way, the plan of the connections of these ganglia with the brain is a far more difficult matter. It is to J. Dejerine,[10] Ramon y Cajal,[11] and more recently to the works of the Vogts, Kinnier Wilson[12] and Ramsay Hunt that we owe such positive knowledge as we possess relative to the fibers which unite the striate body with the adjacent gray matter.

As we have shown above, the caudate nucleus and the external segment of the lenticular nucleus have an identical structure, and so it is legitimate to unite them under a common name—the *striatum* (C. and O. Vogt), which is quite naturally contrasted with that of the globus pallidus—the *pallidum* (C. and O. Vogt).

B. UNION OF THE STRIATUM AND THE PALLIDUM.—The *striatum* (putamen + caudate nucleus) is united to the pallidum (NL^2 + NL^3, globus pallidus) by fine fibers which represent the axis-cylinders of the cells whose character we have described. These fibers come from both the caudate nucleus and the putamen, radiate inwardly and downwardly, crossing the bundles of large fibers which form the internal capsules, and finally reach the pallidum.

The researches of K. Wilson[12] show that the destruction of the putamen in the cercopithecus caused a degeneration of those strio-pallidal fibers, and, moreover, that this degeneration did not pass beyond the pallidum.

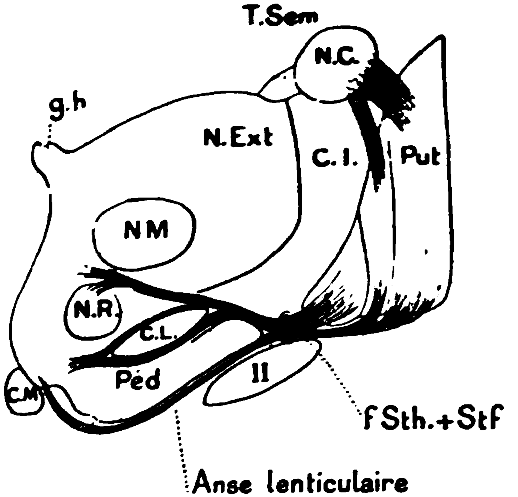

Fig. 1. Diagram of the most important bundles of fibers uniting the various segments of the corpus striatum to each other and to the thalamus. Vertical section of the thalamus, the lenticular nucleus divided into its three segments and the caudate nucleus.

Put = putamen (external segment of the lenticular nucleus NL^3). C.I. = internal capsule; N. Ext. = external nucleus of the thalamus; N.M. = median nucleus; N.R. = red nucleus; C.L. = body of Luys; C.M. = mammillary body; Ped. = foot of the cerebral peduncle; T. Sem. = taenia semicircularis; F Sth. + St.F. = bundles making up the ansa lenticularis which unites the lenticular nucleus with the thalamus, red nucleus, body of Luys and the gray substance of the third ventricle. II = the optic tract.

The question of the connection between the *striatum* and the cerebral cortex was in doubt until recent years. To-day it seems definitely established that there is no *direct* connection, that there are no fibers which unite the cerebral cortex and the striatum. The findings of Economo[13] in a case of destruction of the lenticular nucleus in man, of degenerated fibers radiating in fan-shape in the corona radiata and ending in the precentral gyrus and in the parietal convolutions, were probably due to a concomitant lesion of the optic thalamus. (Fig. 1.)

C. UNION OF THE PALLIDUM WITH THE THALAMUS AND THE HYPOTHALAMIC CENTERS.—The pallidum is united with the optic thalami as well as with the hypothalamic region by very important tracts all of which constitute the *efferent lenticular system*. This lenticular system is, from all the evidence at hand, very old in the phylogenetic sense. It is found in fish and loses none of its importance in mammals and man. Moreover, the time of myelinization of the ansa lenticularis is very early, as Flechsig demonstrated. This ansa lenticularis is quite a complicated system whose elements have been dissociated and described, thanks to the researches of Flechsig, J. Dejerine, Ramon y Cajal, K. Wilson, C. and O. Vogt and Grünstein[14].

Three important tracts appear in the frontal section of the cerebral hemisphere prepared by the Weigert-Pal method. These radiate from the *pallidum*, proceed to the *thalamus* and the hypothalamic region. The most important of these leaves the *pallidum*, crosses the internal capsule and ends in the external lamella of the *thalamus* as well as in the oroventral region of this ganglion. Below this is still another group of fibers which encircles the foot of the cerebral peduncles, gaining the hypothalamic region. In addition to the latter there are fibers arranged in fasciculi, which proceed to the hypothalamic body (body of Luys) and to its capsule.

The terminations of these different bundles have been determined by the secondary degeneration method. This has revealed, among other things, that numerous fibers leave the pallidum, penetrate the hypothalamic region and reach the cerebral peduncle in order to go to the red nucleus and its capsule, to the *substantia nigra,* to the floor of the third ventricle and the *tuber cinereum.* There are others which decussate by means of the ventral commissure of Meynert and go to the opposite hypothalamic region, and to the contra-lateral lenticular nucleus.

Experimentally many authors have been able, by destruction of the lenticular nucleus (*putamen* + *pallidum*) to provoke secondary degenerations of the lenticular system and follow them in the centers above indicated. In dogs and rabbits, Grünstein found three pallidofugal bundles, one ending in the thalamus, another in the *tuber cinereum,* and the last in the body of Luys and the *substantia nigra.*

Muskens was able after the destruction of the lenticular nucleus in a cat to follow degeneration into the external lamella of the thalamus, into the field of Forel, and into the anterior region of the red nucleus. In the cercopithecus, Kinnier Wilson distinguished fine fibers caudally situated and medium sized fibers orally situated. The first proceed to the ventral region of the median nucleus of the *thalamus,* the second end in the capsule of the homolateral and heterolateral red nucleus via the commissure of Forel, in the homolateral body of Luys and in the homolateral *substantia nigra.* According to Wilson, the commissure of Meynert contains no pallido-fugal fibers and only degenerates when it is directly injured.

Let us finally add that C. and O. Vogt were able to follow a bundle which, after issuing from the ansa lenticularis and crossing the capsule of the red nucleus, ended in the nucleus of Darkschewitsch and the nucleus of the posterior commissure. On account of the close relations which unite these nuclei and the posterior longitudinal bundles one may judge the physiological importance which should be attributed to the strio-tegmental bundle of C. and O. Vogt.

To summarize: The pallido-fugal fibers are broken up into bundles which according to C. and O. Vogt may be classified as follows:

(a) *Pallido-thalamic fibers* proceeding to the oro-ventro-medial region of the thalamus.

(b) *Pallido-mesencephalic fibers* (strio-tegmental) for the nucleus of Darkschewitsch and the nucleus of the posterior commissure.

(c) *Pallido-rubral fibers* for the red nucleus.

(d) *Pallido-Luysian fibers* for the homolateral hypothalamic body.

(e) *Pallido-nigral fibers* for the homolateral substantia nigra.

D. UNION OF THE OPTIC THALAMUS WITH THE STRIATE BODY. STRIO-PETAL SYSTEM.—As we indicated above, the striate body (striatum + pallidum) receives no fibers from the cerebral cortex; nor does it receive any from the cerebellum, the red nucleus, the body of Luys, the substantia nigra or the ribbon of Reil. The only actually identified strio-petal fibers come exclusively from the *thalamus.* These are specifically mentioned by J. Dejerine (see the case of Gardette), by G. Roussy[15] in man, and were found in experimental lesions by G. Roussy, Sachs,[16] Pfeiffer,[17] and Kinnier Wilson. These researches show that destruction of the oro-medial region of the *thalamus* causes secondary degeneration of fibers going to the hypothalamic region, the globus pallidus, and ending in the putamen and even in the caudate nucleus (Sachs).

From this account, it appears that the striate body, completely independent of the cerebral cortex, forms a system *anatomically* autonomous, whose *afferent* (strio-petal) fibers have their origin in the thalamus and whose *efferent* (strio-fugal) fibers, arising almost exclusively in the pallidum, proceed to the thalamus and the hypothalamic centers, the *body of Luys,* the *red nucleus,* the *substantia nigra,* the *tuber cinereum* and the region of the cerebral peduncles.

Having no direct connection with the central sensory tracts, the striate body seems, from the anatomical point of view, to be solely a motor center the excitation or destruction of which produces disturbances limited to motor functions. And as a matter of fact, this is what the most recent and best proved literature teaches us.

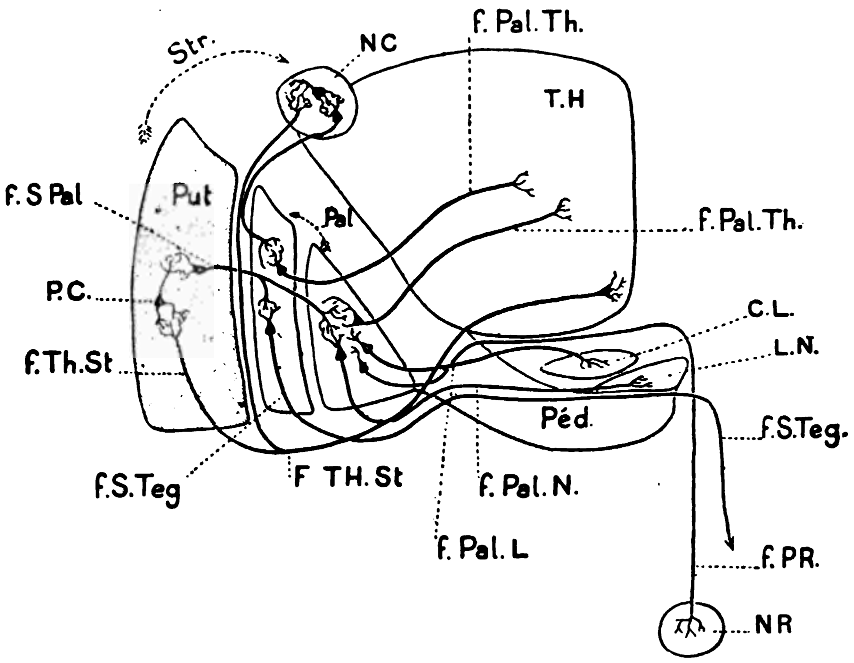

Fig. 2. Diagram of the connections between the thalamus and the corpus striatum and between the hypothalamus and the corpus striatum. Vertical section showing the caudate nucleus (N.C.); the lenticular nucleus divided into three parts, the external being the putamen (Put); the two internal, the globus pallidus (Pal); the thalamus (TH); the hypothalamic centers, the body of Luys (C.L.); the substantia nigra (L.N.); and the foot of the cerebral peduncle (Ped); the *Striatum* (NC + Put) has been shaded, while the *Pallidum* (Pal) has been left white.

i. The striofugal system made up of (a) strio-tegmental fibers (FS. Teg); (b) the pallido-rubral fibers (F.PR.) going to the red nucleus (N.R.); (c) the pallido-nigral fibers (F. Pal.N) going to the substantia nigra (L.N.); (d) the pallido-Luysian fibers (F. Pal.L) going to the body of Luys (C.L.) and the pallido-thalamic fibers (the two bundles of Forel) (F. Pal. Th.).

ii. The strio-pallidal system made up of fibers uniting the *Striatum* and the *Pallidum* (F.S.Pal.); the putamino-pallidal fibers uniting the caudate nucleus with the lenticular nucleus.

iii. The intra-striate system formed by neurones interposed between the striopetal and striofugal neurones.

iv. The thalamo-striate system made up of numerous fibers (represented as one in the figure) which come from the ventro-median region of the thalamus and go to the putamen and the caudate nucleus.

The very suggestive microscopical-anatomical researches of Malone have added an argument of great value. Malone began a systematic study of the encephalo-medullary axis with the idea which is being confirmed each day—that the functions of the nervous system are reflected in their architecture and that nerve centers and nerve cells having different functions will present equally different structure. He was able to show that the *pallidum* was composed of cells of *motor type,* while the *thalamus* contained none of these. Malone also considered that the *globus pallidus* was the essentially motor organ of the lenticular nucleus.

Though anatomy indicates the motor rôle of the striate system, work dealing with its physiology is less precise, as was mentioned earlier in the article. The researches of Magendie, Schiff and Nothnagel, to be sure, seem to show that the striate body possessed a manifest influence on muscular function, but the interpretation of their experiments was soon shown to be questionable because of the associated lesions which, of necessity, resulted from insufficiently delicate technique. It is necessary to recall how extremely difficult it is to produce irritation of the caudate and lenticular nuclei in mammals without there being simultaneous and similar manifestations due to involvement of other structures.

Other experimenters have tried to obtain more definite results, but mention of the conclusions of the most important of the works on this subject is sufficient to indicate their weakness. Pruss[18] by exciting the caudal region of the caudate nucleus, obtained tonic muscular contractions, whereas excitation of the middle region provoked clonic contractions, and that of the anal region movements of locomotion.

In order to avoid the effects of excitation through the capsular fibers (motor cortico-bulbar and cortico-spinal fasciculi) Bechterew[19] removed the motor cortex, and after the pyramidal degeneration was complete, excited the caudate nucleus; no motor phenomena could be obtained, but excitation of the lenticular nucleus provoked muscular contraction. Following an analogous method. Ziehen[20] obtained different results. After removing the cortex of a rabbit, not only was the caudate nucleus inexcitable, but also the entire lenticular nucleus. Minor[21] obtained the same results; according to him, the striate body is not responsive to any excitation.

It is useless to continue such deceptive analysis of physiological works, for the results of Nothnagel, Carville and Duret, Johannsen, Baginski and Lehman, Sgobbo, Sellier and Verger and Lo Monaco will not enlighten us on the subject of the functions of the striate body. We cannot but agree with Luciani[22] that our knowledge of the physiology of the lenticular nuclei is most rudimentary.

III. ANATOMICO-CLINICAL SYNDROMES

A. INTRODUCTION.—The dearth of convincing physiological works on the functions of the striate body obliges us, as said before, to turn to clinical facts, controlled by careful pathological examinations in order to ascertain at least a part of these functions. Here the ground is solid, for works on this subject are not lacking. In fact they are so numerous that a complete analysis would overflow the limits of this article. Therefore only the most significant will be reported.

The anatomy and histology of the striate body have already warranted the suggestion that, owing to their profound architectural and cytological differences, the *pallidum* (globus pallidus, $NL^1 + NL^2$) and the *striatum* (putamen + caudate nucleus) are not identical in function. This hypothesis seems to be well confirmed by all the facts which have been carefully studied during recent years. Moreover, it has now become possible to describe in different chapters a syndrome of the striatum, a syndrome of the pallidum, as well as a syndrome of the entire striate body.

We shall first consider the total striate syndrome, for despite the complexity and entanglement of the symptoms which enter into it, it includes all the symptomatological elements of both the striate and pallidal syndromes.

B. SYNDROME OF THE CORPUS STRIATUM CAUSED BY THE DESTRUCTION OF THE PALLIDUM AND STRIATUM. 1. *The progressive lenticular degeneration of Kinnier Wilson.*—This syndrome, the first observations of which were successively published by Homen[23], Ormerod[24], Gowers[25], and Anton[26], was definitely established by

Kinnier Wilson. He stated its limits precisely and showed the close relation between destruction of the striate body and changes in the liver. Immediately, and quite justly, "Progressive Lenticular Degeneration" was called "Wilson's Disease."

The first symptoms of the disease always appear between the ages of ten and twenty-six. Even though no basis for the disease can be traced in the antecedents of the patient, it is striking that it often assumes a familial character and most frequently strikes large families.

One of the earliest manifestations of progressive lenticular degeneration is the appearance of constant involuntary movements consisting of a true and rhythmical tremor of the upper extremities, causing contraction of the antagonists. The rate of this is from four to eight oscillations a minute. Attention and voluntary efforts, as well as psychic or sensory stimuli augment its intensity. During voluntary movements which call for effort, the tremor disappears in the active member but appears in the unaffected one due to the dispersion of the excitation. Limited at first to the upper extremities and subject to remissions, the tremor spreads to the lower extremities and finally to the head. As Wilson pointed out, muscular relaxation as well as sleep causes the tremor to disappear.

Besides these rapid and rhythmic contractions, transitory contractions are often found which cause special attitudes of the extremities. Frequently the arms extend or flex by means of a slow contraction of the extensors or flexors, then undergo an inward or outward twisting, the fingers also presenting movements analogous to or identical with those of *athetosis.*

It is not necessary to emphasize how inconvenient this tremor and these athetoid spasms which we have just mentioned may be to the exercise of the functions of the extremities. But the most conspicuous and constant hindrance of these functions is without doubt, *muscular rigidity.* This symptom may be considered the most important of Wilson's disease.

Starting with the lower extremities, the hypertonia extends to the upper extremities, invades the neck and face and finally the trunk. The entire skeleton is thus held by tense musculature which limits active as well as passive movements. Held in a general attitude of flexion, the patient carries out even the most elementary movements with great slowness and difficulty. If one attempts passively to displace any part, the hypertonia appears more clearly than ever, the antagonistic muscles become taut beneath the skin and may be seen in full outline. It appears as though the muscles were transformed into bundles the consistency and elasticity of which resemble that of rubber.

The muscular rigidity does not spare the muscles of the face and one is often, even at the beginning of the disease, struck by the facies it causes. Immobile, sad, the mask has lost all expression and recalls that of an imbecile or even an idiot in whom no emotion comes to modify the immovable and frozen features. Only the eyes, whose mobility and expression are preserved attest that behind the inexpressive mask active psychological processes are going on. Occasionally a vague beatific smile appears and seems like a wave of life passing over a frozen face. But the monotony of the expression as well as its limitation shows that it corresponds to no real sentiment and is, in truth, only due to fixation of the lower type. In addition to the defect of facial mimicry it is interesting to note the appearance of *massive emotional movements* which pass across the habitually immobile features. By this we mean the fits of *crying* and especially of *spasmodic laughter* whose unexpected coming is not in any way linked to the appearance in conciousness of an adequate affective stimulus.

The hypertonia, which fixes the limbs, trunk and face in attitudes which are hard to modify, does not spare the muscles which concern speech, phonation or swallowing. Hesitating, heavy, occasionally nasal, speech soon becomes incomprehensible, and it is not unusual to find the unfortunate patients unable to express themselves save by monotonous grunting. Both anarthria and aphonia are absolute in the most complete sense. Difficulties in swallowing are also the rule, and in some cases dysphagia is so great that fluids are expelled through the nose. As far as can be determined by a pharyngo-laryngoscopic examination, these very profound modifications of phonation are not in any way related to a paralysis of the muscles which are responsible for its functions. Slowness of contraction of the palato-pharyngeal muscles, a "dragging" of a vocal cord in closing of the glottis, may be found.

As Wilson pointed out, though rigidity, tremors and spasms constitute the principal obstacles to the accomplishment of muscular functions, there is also another element to be considered—asthenia. This is shown by the inability of the patient to prolong the slightest effort. Neither closure of the eyelids nor projection of the tongue, can be maintained more than a short second. It is also noteworthy, that cases of progressive lenticular degeneration present a most striking state of motor apathy. The patients, inert in their chairs or beds, do not seem to concern themselves with anything or attempt to make any voluntary movements. If they wish to satisfy their needs, they get up slowly and then stand stupidly and as if frozen, without carrying out the act which they had seemingly determined upon. (Raymond and Lejonne). In some cases voluntary movements are not only interrupted by spasms but also by wild gestures, brusque, jerky, truly *choreiform.*

We cannot dwell on the "negative symptoms" of Wilson's disease without the risk of prolonging this account beyond reason. However, the absence of all "pyramidal" involvement (Babinski's sign, absence of the normal skin reflexes, paralysis), of any change in trophic function, forms a sufficiently striking and characteristic contrast to the positive striatal signs which have been emphasized.

Wilson, as well as the early observers of progressive lenticular degeneration, mentions profound psychic disturbances in addition to those of motor functions. The cases observed by Gowers and Ormerod presented torpor and emotional indifference truly pathological, and occasionally a diminution of memory. But the intelligence of the patients remained without noticeable impairment. Beside this most frequent form of psycopathy, there is another whose characteristics Wilson has described. This is not caused by a benumbing of the faculties, but on the contrary, by psychic *excitation.* Always full of life, amused, the patient shows a perpetual gaiety which forms a sad contrast to the physical impairment.

Wilson's disease presents equally definite anatomical characteristics. Horizontal sections of the hemispheres show the fundamental lesion at first glance—extreme atrophy of the *striatum* and *pallidum.* Moreover, foci of more or less extensive degeneration appear in the *putamen.* This, as is shown microscopically, is composed of granular cells surrounding undiseased blood vessels. In cases where the disease developed more slowly, the nucleus has entirely disappeared, being replaced by a cystic cavity. The internal capsule and the *thalamus* are intact; the only secondary degenerations to be found are limited to the efferent striate fibers—striothalamic and hypothalamic fibers (see chapter on anatomy). The ansa lenticularis always appears very much reduced in size. The fact that it is never entirely degenerated is due to the strio-petal fibers of thalamic origin which it contains.

Besides these striate lesions others may be found, as was shown by Lejonne and Lhermitte[27] in those cases in which psychic as well as striate symptoms existed. Volumetric reduction of the corpus callosum, diffuse demyelinization of the corona radiata and atrophy with increase of the lipochrome of the cortical cells have been observed.

One of the anatomical peculiarities of progressive lenticular degeneration consists of marked and peculiar involvement of the liver. Wilson pointed out that the moderate atrophy of the liver is accompanied by a very apparent deformity of its surface which appears greatly modified. It is hobnailed by the retraction of scar tissue. The intrahepatic development of connective tissue is accompanied by profound alterations of the glandular elements of which the most characteristic are nodules of hyperplasia, steatosis and necrosis. In addition to these hepatic lesions others are occasionally found: sclerosis of the splenic pulp and of the pancreas with thickening of the vascular walls of these organs (Lhermitte[28]), calcification and sclerosis of the aorta and large vessels, and renal sclerosis. All of this shows how diffuse the cause of lenticular degeneration may be.

2. *Pseudo-sclerosis.*—Another malady, first described by Westphal[29] and then by Strümpell[30] under the name of *pseudo-sclerosis* is clinically, at least, very similar to Wilson's disease.

This disease has certain of the symptoms of Wilson's disease with its slackening of voluntary movements, the dysarthria, the slight paresis of the extremities, the tremors which are rendered more apparent by concentration of mind or emotion, and finally the psychic disorders—disorders of character, contraction of the intellectual horizon, and diminution of attention. Changes of the liver and the spleen are found in Westphal-Strümpell[30] disease as well as in progressive lenticular degeneration; they are apparent both clinically and histologically. One of the peculiarities of pseudo-sclerosis is that it is accompanied by a special type of pigmentation of the periphery of the cornea. The sclero-corneal limbus, Descemet's membrane and the vitreous assume a brownish-green tinge which presents, as Fleischer[31] and A. Rumpel[32] pointed out, many of the characteristics of the pigmentation of argyria.

That which very neatly differentiates pseudo-sclerosis and which we think prevents one from considering it a *forme fruste,* a sketch of Wilson's lenticular degeneration, is the group of lesions which cause it. The alterations of the encephalon not only appear less obviously but they require appropriate and special technique to make them appear at all. They consist, as Alzheimer[33] showed, of a proliferation of the fibrillary neuroglia which extends into the white substance of the hemispheres, and involves with striking predominance the striate bodies. But this is not all. In the lenticular and caudate nuclei, the microscope shows on one hand an augmentation of the normal neuroglial stroma and on the other neurological elements unlike any of the normal types. These are, among others, giant cells deprived of fibrils and with much reduced protoplasmic elements, in the center of which is a large sharply defined nucleus. These findings by comparison with teratological processes, suggest the hypothesis, not actually verified, that pseudo-sclerosis belongs in the large group of dysembryo-blastomatoses.

3. *The Senile and Presenile Pseudo-bulbar Paralysis Syndrome of Striate Origin.*—The symptomology of Wilson's disease seems at the onset so striking that its resemblance to the classical picture of senile pseudo-bulbar paralysis could not fail to strike the minds of the first observers. And this actually occurred as is shown by the number of observations on progressive lenticular degeneration published under the title of "pseudo-bulbar paralysis" in children.

It is necessary to dwell on the points of similarity between these two types of disease and to recall that in senile pseudo-bulbar cases the following are found as in lenticular degeneration of adolescence: uncontrollable crises of crying and laughing, dysarthria and aphonia, dysphagia with the return of liquids through the nose, loss of facial and gesticulatory mimicry, voluntary akinesia or loss of spontaneous movements, disappearance of associated movements, which are among the most significant components of primitive automatism, hypertonia with slackening of voluntary movements and a set attitude of the trunk and head in semi-flexion. The expression of beatific stupidity or of emotional indifference was not the only indication of the close relationship of the two syndromes which we have described.

Nevertheless there are certain manifestations which belong almost entirely to Wilson's disease, and when these are added to the pseudo-bulbar syndrome, they present an unmistakable picture of lenticular degeneration. We mean by this the spasms which fix the limbs in attitudes of extension and tension, the tremor, and especially, the movements of a choreo-athetoid type.

However important these last phenomena may be from the point of view of the meaning of the syndrome, they must not efface the resemblance which we have found so marked between Wilson's disease and certain types of pseudo-bulbar paralysis in adults as well as in the aged.*

Moreover, the resemblance does not end with the clinical characteristics but extends to the types and to the topography of the anatomical changes. Wilson's disease is characterized by volumetric reduction of the striate body and a neuroglial sclerosis in the lenticular and caudate nuclei, and, in cases which progress slowly, by a massive destruction of the *putamen* and caudate nucleus; but there is a form of senile pseudo-bulbar paralysis which has as its fundamental anatomical characteristic a generation of the nerve elements of the *striatum* and of the *pallidum* (*status desintegrationis* of C. and O. Vogt). This is manifested to the naked eye by small lacunar areas of softening, a mottled appearance and yellowish cysts which give evidence of old hemorrhages. Such distinctive lesions cause, it is thought, secondary degeneration not only of the strio-pallidal fibers but also of the pallido-fugal thalamic and sub-thalamic fibers (pallido-nigral, rubral, tegmental, etc). However we must insist on the fact that even though the lesions in both Wilson's and pseudo-bulbar palsy are dependent upon a *destructive* process, this destructive process is different in these two diseases. In pseudo-bulbar palsy the destruction has its origin in the grossest kinds of vascular alterations; in Wilson's disease, even though the loss of nerve and neuroglial elements have a paravascular origin, the lumen of the capillaries, veins and arteries remain quite normal. Progressive lenticular degeneration depends upon a toxic necrosis, while the lenticular degeneration of old age is due to an anoxemic necrosis.

4. *Striate syndromes caused by presenile lesions of the striatum and pallidum. The senile Parkinsonian syndrome* (C. and O. Vogt).—The most important elements of the syndrome are common to it and those which have preceded it. That is to say that its characteristics are concentrated in the domain of those automatic movements which have to do with the gait, the mimicry of gestures and of the facies, with phonation and deglutition. Furthermore, muscular tone is increased without the hypertonia showing any of the classic signs of spasticity of pyramidal origin.

Just as in cases of pseudo-bulbar palsy, the gait is slow with small steps and interfered with, at times, by irresistible pulsion. Brachybasia, general rigidity of the half-flexed trunk, pulsion, expressionless appearance of the face and slowing of voluntary movements are the characteristics which lead one's thoughts to the Parkinsonian syndrome. They permit the inclusion of this type among the Parkinsonianisms. Or it may be called the Parkinsonian syndrome if one agrees with those authors who refuse to consider paralysis agitans as a separate disease. (J. Ramsay Hunt, C. and O. Vogt, Mott[34].)

We should like to emphasize that the syndrome which we have just described is not accompanied by tremor and that if it is to be included in the Parkinsonian syndrome it must be admitted that it is a special form of the disease, *paralysis agitans sine agitatione*. Furthermore, contrary to the classic form of paralysis agitans, this form is accompanied by special symptoms, notably torsion spasm and athetoid movements. These motor manifestations are so important that, as has been observed by Thomalla[35], they mark the beginning of striate disintegration.

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* It is worth mentioning that besides the pseudo-bulbar paralysis resultant upon striate lesions there is another type caused by bilateral degeneration of the cortico-bulbar and cortico-pontile tracts.

The anatomical basis of the syndrome rests upon the observations of Westphal and the Vogts who have shown that there exist neuroglial proliferation in the striate, calcareous infiltration of the striate vessels and accumulation in the lenticular nucleus of globular masses of a substance which has not yet been chemically identified (G. Perusini).

C. SYNDROMES OF THE STRIATUM (caudate nucleus + putamen, third segment of the lenticular nucleus).—As we have already pointed out, the anatomical evidence is perfectly clear that the corpus striatum may be resolved into two organs which are histologically quite distinct; the *striatum* (putamen and caudate nucleus), and the *pallidum* (two internal segments of the lenticular nucleus, or globus pallidus).

Keeping to the fundamental principle that should guide all neuro-pathological and neuro-physiological studies, namely, that an anatomical differentiation should be accompanied by a functional one, we can see that lesions strictly limited to the *striatum* or to the *pallidum* cannot be expressed by the same terms—that is, the symptomatology of the one cannot be identical with that of the other.

This hypothesis is amply justified by the anatomical-clinical facts which show that alongside of the total striate syndrome (complete necrosis of the lenticular and caudate nuclei), there is room for a syndrome of the *striatum* and a syndrome of the *pallidum*. Yet it would be incautious pathology to assume that every destructive lesion of the striatum should be translated into an identical syndrome. In fact, that would be reckoning without the double influence of the lesion itself and its point in time. One need only recall the trite but true observation, that the child's reaction to a lesion of the nervous system is quite different from the adult's, and that an alteration of the same character in the same location could assert itself very differently in the two instances. If any one should doubt this, the facts which we are to analyze will offer striking proof of its correctness.

1. *The syndrome of Cecile Vogt, with marbled appearance of the striatum. Little's disease, with congenital and regressive spasticity alone.*—Although the different clinical types of spastic rigidity of the lower extremities of the new-born have been known for a long time, and Pierre Marie[36] clearly distinguished two types of infantile cerebral diplegia in 1886, one with and the other without pyramidal degeneration, it must be recognized that we owe the anatomical-clinical individualization of this complex to the work of Cecile Vogt, and that it is this author who first arranged a well defined grouping of the symptomatic elements in the striate syndrome. The syndrome of C. Vogt is always characterized by bilateral motor disturbances. The most important consists in the hypertonicity of striated muscle, the slowing, awkwardness, sometimes temporary impossibility, of voluntary movements, quite as marked in the upper extremities as in the lower. This spasticity is free of all paralytic components. Sometimes, as in a case of Freund's, the hypertonicity shows itself more in the lower limbs, so that one could almost describe a paraplegic form of this syndrome with a springing gait.

Marbling of the striatum is represented not only by general or selective spasticity, but also by involuntary movements, constantly unstable station, and especially by *bilateral athetosis*. To the latter are added associated movements which may suddenly exaggerate the disturbances of voluntary motion. At times there are attacks of weeping or spasmodic laughter. Attention, concentration upon a volitional movement, external stimuli, and emotions, all serve to exaggerate the involuntary movements as well as the spasmodic laughter and weeping. As for voluntary movements, they are greatly diminished. The syndrome of C. Vogt is not accompanied by any sign belonging to the pyramidal or cerebellar systems. One would search in vain for a modification of the tendon, bone, or skin reflexes, and the great toe sign (Babinski's) is absent. On this last point, however, there may be some uncertainty which should be dispelled.

Babinski's sign consists, as is well known, in extension (dorsiflexion) of the great toe following plantar stimulation. Now, in several cases of Vogt's syndrome, not only has spontaneous extension of the great toe been observed, but an inconstant occurrence of this movement has *apparently followed* plantar stimulation. In one of her earliest studies, C. Vogt was in doubt as to whether this variable and uncertain extension of the great toe should be deemed a true Babinski, or whether it might not be preferable to assume a *pseudo-Babinski* in addition to the true phenomenon, and was inclined to prefer the latter interpretation. There is no hesitation to-day, and C. and O. Vogt have explicitly stated in their latest contribution that lesions of the striate body cannot induce the reflex extension of the great toe, the genuine Babinski, but that when such an extension is occasioned in the striate syndromes it is no more than an expression of athetosis. An identical conclusion was reached by Lhermitte and Cornil[37] in their study of a striate syndrome in adults.

In its evolution, the syndrome of C. Vogt presents one fundamental characteristic, regression or subsidence of symptoms, though this is by no means equally marked in all cases.

On the anatomical side, the marbled appearance is brought about by a reduction in volume of the striatum, associated with a peculiar alteration consisting of the replacement of striate cells by a packing of fine myelinated fibers, absent under normal conditions. It is exactly this fantastic arrangement of myelinated-fiber plaques, brought out by the Pal method, that produces the marbling of the striatum.

2. *Dysmyelinization* (C. and O. Vogt). *General rigidity with progressive terminal athetosis.*—This anatomical-clinical syndrome is really closely connected with the preceding one. What separates them is, on the one hand, the difference in development, progressive in the dysmyelinization and retrogressive in the marbling, and on the other hand, the different periods for their appearance. The former is an acquired condition, the latter congenital.

The condition of dysmyelinization generally makes its appearance in the first months of life (cases 1 and 2 of C. and O. Vogt), but may not do so until adolescence, as evidenced by the observations of Rothmann and O. Fischer. The cardinal signs of this syndrome, rigidity subject to temporary exacerbations, and athetosis, recall very clearly those of marbling.

Anatomical studies demonstrated to C. and O. Vogt that it was a question of degeneration of the strio-pallidal and pallido-thalamic fibers.

This anatomical-clinical type approaches the *dystonia lenticularis* of which Spiller[38] has recently reported a typical example. The latter affection develops in the guise of an acquired double athetosis and ends its evolution with the termination of life after some years. As in Wilson's disease, with which it has a closer bond of relationship, dystonia lenticularis is accompanied by profound difficulties in the articulation of words and in deglutition. Anatomically, the most striking modification consists of the very marked breaking down of the putamen and the caudate nucleus, associated with a reduction in size of the *globus pallidus* without change in its fibers.

3. *Huntington's chronic progressive chorea, result of "atrophic cortico-striate degeneration"* (Pierre Marie and J. Lhermitte[39]).—Since Anton's fundamental observation of the intimate dependent relationships that connect choreic and athetoid movements with the striate body, the attention of anatomists has been drawn to lesions of the nuclei at the base of the brain in the choreiform affections. Since the work of Jelgersma[40], Alzheimer[41], Kleist[42], Pierre Marie and Lhermitte, Kiesselbach[43], and C. and O. Vogt, the problem of the anatomical origin of chorea may be considered solved.

In a short note, Jelgersma showed that chorea of the Huntington type was accompanied by an atrophy of the caudate nucleus in its anterior portion especially; then Alzheimer, in three cases, demonstrated degeneration of the nerve cells of the caudate and lenticular nuclei, accompanied by extreme proliferation of the neuroglial web. Analogous lesions were discovered by Kleist in a case of chronic chorea that had progressed for fifteen years. Here again the neuroglial sclerosis was particularly severe in the striate body, the red nucleus, and Luy's body (nucleus hypothalamicus); and certain regions of the thalamus as well exhibited degenerative changes.

Taking up this anatomical study in four cases of typical Huntington's chorea, Pierre Marie and Lhermitte established the following: (1) Considerable gross atrophy of the lenticular and caudate nuclei; (2) very profound degenerative changes in the nervous elements, cells and fibers of these ganglia; (3) an intense reaction of the neuroglia in the same regions; (4) chronic changes in the vessels of the striate body, and (5) an atrophy of the frontal-Rolandic cortex without secondary degeneration of the cortical projection-fibers. These authors insisted on this point, that not only are the changes unrelated to the thalamus and to the centers beneath the striate body, but they display a striking predilection for the putamen and the caudate nucleus (striatum). In the system of the ansa lenticularis the bundles from the striate to Luy's body and to the red nucleus were only reduced in size, not degenerated, which is explained by the preservation of the centers of fibers efferent from the pallidum.

Though the neuroglial proliferation constitutes from all the evidence an important feature of the lesions in chronic chorea, it is far from essential.

Lhermitte and R. Porack[44] have shown, in fact, that the neuroglial sclerosis may be entirely lacking, in spite of intensity of degeneration in the striate body. Kiesselbach, Dunlap[45], and C. and O. Vogt have found these alterations in all the cases of chronic chorea that they have been able to study, and C. and O. Vogt have reported further a degeneration of the striatum-to-pallidum fibers, and in some cases a thickening (hydropic degeneration?) of the large myelinated fibers of the external segment of the pallidum.

The cortico-striate atrophic degeneration is represented by the chronic progressive chorea of Huntington, but there is reason to believe that all the chronic choreas imply lesions over the same distribution at least, if not of an identical character.

The muscular disorder of Huntington's hereditary chorea resembles, in certain of its traits, that provoked by the complete necrosis of the striate body (striatum + pallidum); and it has long been recognized that involuntary movements are very often a mixture of chorea and athetosis, so much so that in certain subjects it is extremely difficult to decide whether it is a condition of true chorea or of athetosis. Yet there is one characteristic, lacking in Huntington's disease, that on the contrary appears constantly in the lenticular degeneration of Wilson, namely hypertonicity.

Not only is muscular tone not exaggerated in chronic chorea, but it is even clearly diminished, as Lhermitte and Lamaze[46] have shown recently.In certain cases this hypotonicity is as striking as hypertonicity may be in Wilson's disease.

In the last-named affection, voluntary movements are diminished, and alternating, diadochokinetic movements are particularly difficult. According to C. and O. Vogt, it would be impracticable to distinguish this disturbance of innervation from the true adiadochokinesis encountered in lesions of the cerebellar system (Babinski), and one is justified in considering it as they do, as a pseudo-adiadochokinesis. In a typical case of Huntington's chronic chorea, Lhermitte and Lamaze recently called attention to the slowness and difficulty of alternative movements, and asked whether it were not a question of true adiadochokinesis, since in this case the hypertonicity with the muscular rigidity which it determines was totally lacking. It was the same in another observation, of unilateral syndrome of the striatum, reported by Lhermitte and L. Cornil, where adiadochokinesis was evident in spite of the absence of hypertonicity. Regarding this very important point, it would be hazardous indeed to base a conclusion upon so small a number of positive cases not yet anatomically controlled. However, we are greatly inclined to believe that *true* adiadochokinesis is not the exclusive property of cerebellar lesions, but that it should be accorded a place among the striate symptoms.

4. *Acute Lesions of the Striatum. Sydenham's chorea.*—In contrast to the lesions of chronic development, those that represent a toxic-infectious process in the brain affect a limited area only exceptionally. And we do not know to-day whether there exist any encephalitides limited to the striate body. However, we cannot fail to mention the very interesting researches of Pierre Marie and Tretiakoff[47] in Sydenham's acute chorea, and the proof that they gave of alterations especially pronounced in the lenticular and caudate nuclei. True, the symptomatology of acute chorea is too rich for all its elements to find an explanation in lesions exclusively striate, but there is nothing to prevent these lesions being attached to the choreic muscular disorder at least, as well as to the hypotonicity whose existence we recognized in Huntington's chorea.

5. *Lesions (hemorrhages or softening) in the region of the striatum.*—We have already alluded to the observation published in 1895 by Anton[48], which warranted his attaching the choreo-athetoid disorder to the striate body.

Recently C. and O. Vogt have been able to study a case where a softening with cystic transformation of the head of the caudate nucleus and of the adjacent portion of the putamen was expressed by a contralateral hemichorea.

In an observation of Lhermitte and Cornil cited above, a stroke in a woman of fifty had determined a hemichorea-athetosis with spontaneous clonus of the foot and pseudo-Babinski, independent of any paralysis or hypertonicity.

These several facts demonstrate that *unilateral* destructive lesions of the striatum do not remain silent though sometimes sparing of symptomatology. The involvement of these centers reveals the coordinator rôle of the striate body.

D. Syndromes of the Pallidum.—By definition the pallidal syndrome should be the clinical expression of lesions strictly limited to the globus pallidus, that is to say to the two internal segments of the lenticular nucleus. As a part of this organ Ramsay Hunt includes the large-celled neurones which as we have seen above (see anatomical chapter) are scattered over the putamen and the caudate nucleus (type 1 cells of Golgi, motor type cells of Malone). Following the American neurologist, the pallidal syndrome would appear to be determined not only by destructive or degenerative alterations affecting the globus pallidus, but also by those that reach the pallidal element carried to the interior of the striatum along with the purely striate neurones ("strio-pallidal" system).

This anatomical information, indispensable to an understanding of the facts, accounts for many of the obscurities and contradictions that appear to the reader of works bearing on this subject. In the pallidal syndrome, in fact, we come upon a territory less known than that which we have just explored. It is nothing less than that of Parkinson's disease, and we know all the old and new discussions that this has provoked.

Agreement seems to have been reached on at least one point, of realizing that the Parkinson's disease of the classics cannot be understood as a pure disease entity, possessing as such an unequivocal symptomatology whose lesions have always an identical character and distribution and whose development can be exactly foreseen. This fragmentation of paralysis agitans which is in process of being realized to-day and appears so full of promise has already been productive of fruitful results.

It has been known for a long time that the Parkinsonian syndrome can appear in early life, and that in cases of this kind the disease copies that of old age and is oftenest familial or hereditary. Ramsay Hunt was able to study three cases of precocious "Parkinson's disease" both from the clinical and from the anatomical point of view, and demonstrated in all these cases an identical lesion, very specific in its distribution and character. It is in reality a primary degeneration by abiotrophy of all the pallidal system. That is to say, the cells of Golgi's type I of the striatum, and cells of the globus pallidus are reduced in number and size, a large number have disappeared and consequently the striothalamic and hypothalamic radiations are contracted and atrophied. The gray centers to which these striate radiations extend, the thalamus, substantia nigra, Luy's body, and red nucleus, are untouched.

There is then a juvenile or infantile type of paralysis agitans of Ramsay Hunt, which is induced by an abiotrophy or progressive degeneration of the whole pallidal system.

Is it the same in the paralysis agitans that supervenes at a more advanced age, which one may call *presenile paralysis agitans?* That is the question put by Ramsay Hunt. Before him, Lewi had already localized the alterations of classic Parkinson's disease in the lenticular nucleus and the nucleus of the ansa peduncularis among others, but indications were incomplete. It remained for Hunt to state precisely the anatomical basis of presenile paralysis agitans. In two cases respectively seven and eight years advanced, this author proved by the aid of exact technique the atrophy of the "motor" cells of striatum and pallidum, contrasted with the integrity of the small cells of the striatum.

These lesions mainly affected the anterior regions of the pallidal system. In addition, there was a quantitative reduction of the efferent fibers from the pallidum (thalamic and hypothalamic bundles). As a matter of fact, the circulatory apparatus of the striate body was intact, without a trace of porosity or cribration.

It is impossible not to be struck immediately by the very great similarity (not to say identity) between the striate lesions in the Parkinsonian syndrome of adolescence and that of the presenile period; in the first as in the second, the primary degeneration of pallidal cells stands foremost and incontrovertible. A degenerative abiotrophic process, comparable to that of amyotrophic lateral sclerosis or of the disease of Aran and Duchenne—a selective process confining its effects to a limited anatomical-physiological system—that is the mode of attack of the yet undetermined causative agent, at least of a certain form, of juvenile and presenile paralysis agitans.

Having ascertained this, we immediately understand why these Parkinsonian syndromes, arising in such different periods of life, approach each other to the point of confusion.

But the preceding facts, important as they are, do not exhaust the pathological anatomy of paralysis agitans.

In seven cases studied by C. and O. Vogt, the modifications of the striate body appeared appreciably different in nature. As they note the atrophy of the caudate nucleus accompanied by degeneration of its cells and the fibers emanating from them, so they also find foci of softening, lacunae in the lenticular nucleus, sometimes simply a perivascular disintegration with cavities or their beginnings, with advanced or early porosity.

According to C. and O. Vogt, the lesions of presenile paralysis agitans do not possess a selectivity so pure as in the cases studied by Ramsay Hunt. Those authors acknowledge that there are some cases in which the degeneration appears much more marked in the pallidum, while they claim that the lesions in other cases, without sparing the globus pallidus, do attack the striatum with greater severity. This is not an anatomical statement of merely passing interest; rather it has a far greater range, for C. and O. Vogt remarked that these cases, where the Parkinsonian syndrome was expressed by pronounced trembling, were exactly those which corresponded to the most extensive lesions of the striatum, whereas on the contrary the paralysis agitans without rigidity answered to rather more limited alterations of the pallidum. Can other cephalic centers be involved by the paralysis agitans process, and is it always restricted to the striate body? Apparently this should not be answered in the affirmative. Treitiakoff[49], and Souques and Treitiakoff[50], have certainly shown the frequency of atrophy of the elements of the *substantia nigra* in Parkinson's disease; and if this lesion has not been taken up by Ramsay Hunt nor by C. and O. Vogt, we for our part have never known it to fail macroscopically in four cases of presenile Parkinson's disease which we have had the opportunity of studying. Furthermore, this lesion of the substantia nigra is not surprising, now that we know the close dependence of this mesencephalic center on the striate system. Might there be a variety of neural degeneration illustrated by the crossed hemiatrophies of the cerebellum following lesions of the hemispheres? However that may be, the researches already set forth give us this to remember, that besides the Parkinsonian syndromes of old age conditioned by destructive lesions of vascular origin in the striate body, there exists a type of paralysis agitans of the adolescent or the aged subject which can properly be charged to an abiotrophy with an affinity for the pallidal system.

Progressive degeneration of the pallidum does not seem to assert itself exclusively through the Parkinsonian syndrome, especially when it is associated with a pyramidal degeneration.

We have seen these patients affected with spastic rigidity of all four extremities and the trunk, with the appearance of far advanced pseudo-bulbar cases, swallowing laboriously, unable to utter a sound, essaying a vague pretense of a grin in an occasional spasmodic burst of exasperation, otherwise displaying a facies so completely expressionless and stiffened that it seemed to denote an annihilation of the faculties, yet in whom a more careful examination would demonstrate their intellectual integrity. This clinical picture, of which Lhermitte, Cornil, and Quesnel[51] have reported a striking example, certainly presents very complex features, some allied to those of pseudo-bulbar paralysis, some to amyotrophic lateral sclerosis, some to Parkinson's disease. What differentiates it absolutely from this last is the group of "pyramidal" signs found associated with the striate manifestations proper.

Is it possibly in this group of cases that one should place the curious affection described by Foerster under the name of *arteriosklerotische Muskelstarre* (rigidity)? But at present we lack sufficient descriptions to solve the problem of identifying this "arteriosclerotic rigidity".

IV. SYNTHESIS OF THE SYMPTOMATOLOGY OF PATHOLOGICAL STATES OF THE STRIATUM

After analyzing in the preceding chapters the different symptomatic characteristics of diseases of the striate body, we may now rearrange them more compactly to show their general order and coherence.

First of all we are struck by the fact that the morbid manifestations of the striate body are exclusively *motor*. In the total striate syndrome as well as in the syndromes of the striatum and of the pallidum, all sensory, trophic, and secretory disturbances are lacking. The disorders of muscular function are wholly concerned with voluntary movements, muscle tone, automatic mechanisms of expression, articulation of words, phonation, and deglutition.

One of the fundamental elements of striate symptomatology is unquestionably the *muscular rigidity* of Parkinsonian type, very different from pyramidal spasticity. Indeed we find it quite as much in Wilson's disease and in C. Vogt's syndrome as in paralysis agitans. It is the factor that largely conditions the vicious attitudes of trunk and extremities, and also the slowness of voluntary movements and the pseudo-adiadochokinesis; but it is not the sole cause of irregularities in voluntary motion, since the latter appear at times in those curious cases of Parkinson's disease without rigidity, and in the chronic choreas with hypotonicity. As Van Woerkom[52] has shown very well, what appears to be the essential motor difficulty is the impossibility of quickly substituting the dynamic function of the muscles for their static function, causing an apparent lack of coordination which gives the voluntary movement a "stiff and slow, sometimes an exaggerated, quality."

Furthermore, spontaneous voluntary movements are markedly diminished; Ramsay Hunt and C. and O. Vogt insist strongly on that point. This *general immobility* is especially striking in Wilson's disease and paralysis agitans. But the loss of *normal associated* movements appears no less striking, and with it one can connect up the lack of harmony in movements taken together, the absence of compensatory arm movements during walking, the extreme diminution in movements of defense and of orientation, in fact all the reflex movements that go to make up a harmonious whole in the activities of a normal individual.

All these symptoms, minutely analyzed by Zingerle, are independent of muscular rigidity, as we have already noted how apparent they can be in paralysis agitans without rigidity.

Disturbances of the normal facial expression and gesturing that accompany language and thought naturally fall into the same group. This facial and gesticulatory amimia impresses us, as it does C. and O. Vogt and Hunt, as the most conspicuous and invariable symptomatic peculiarity of the striate syndromes. To it is due the fixed stare, the blank indifferent face, which reflects no *shades of expression,* but explodes in *forced and violent bursts* of spasmodic laughter or weeping.

Finally, the difficulties of lip, tongue, pharynx and larynx motility, which are responsible for dysarthria, anarthria, or even complete aphonia, as well as for the dysphagia that causes an *apparent* exaggeration of salivary secretion in so many pseudo-bulbar and Parkinsonian cases, all these difficulties seem to be related to the preceding group.

Motor manifestations of another order, forming the second great group of striate symptoms, may be placed beside the disturbances of voluntary, normal associated, and automatic coordinated movements. We mean the *spasmodic, choreic,* and *athetoid* types of *involuntary movements.* Different as they appear at first glance, they are none other than links, we believe, in the same chain of variously associated elements. We have already spoken of how the muscular disorder in chronic chorea may be so nearly allied to athetosis that it is sometimes difficult to describe the involuntary movements with one definite label; but there is more than this, and one may conjecture whether the quality of the movement, choreic or athetoid, is not conditioned wholly or in part by the different modifications of muscular tone in each case.

However that may be, all the facts reviewed cannot fail to be extremely instructive from the light that they shed not only on the motor, coordinating, and inhibiting functions of the lenticular and caudate nuclei, but also on different disease types so far removed from each other in appearance. In spite of their superficial dissimilarity, who can fail to be struck by the symptomatological bonds that unite them to form one great pathological group with a changeable countenance like a living face, where the same traits and same wrinkles can be discovered without difficulty under diverse expressions? Whether it be Wilson's disease, C. Vogt's syndrome, pseudo-bulbar paralysis, paralysis agitans, or one of the chronic choreas, we grasp the same fundamental rudiments, mixed in varying proportions and differently arranged, whose origin is placed by pathological anatomy in alterations of the striatum and the pallidum.

What appears most clearly when we leave anatomico-clinical syndromes and attempt to picture the functions of the corpus striatum as a whole, is that the influence of the purely motor functions of this structure are very different from that of the cortico-bulbo-spinal system (pyramidal system). The latter has to do with the most highly differentiated and delicate movements. They may be considered the most intellectual and consequently the most human. All the more rudimentary functions are carried out by the striate system. They are the older movements of the vertebrate series, and, continuing the metaphor, more animal in character.

From the physio-pathological point of view, those conditions which bring about more or less complete destruction of the corpus striatum affect functions which are essentially automatic and which are but very slightly influenced by the cortex. Walking, the synkinetic movements of defence and of spatial orientation, the mimicry of gesture and facial expression, swallowing and phonation are all functions of an automatic character. It is these automatic functions, long gone from the plane of consciousness, which, as the Vogts pointed out, are affected by disease of the corpus striatum.

Everything leads us to the general conception that the motor corpus striatum and the sensory optic thalamus correspond to the precentral motor gyrus and the post-central sensory gyrus which receives conscious sensations. And, further, the corpus striatum and the optic thalamus control the superior automatic motor system, while the cortical centers dominate and control the voluntary motor system.

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THE CLINICAL PHENOMENA OF DECEREBRATE RIGIDITY

WITH A CASE EXHIBITING SOME OF THE SYMPTOMS OF THE CONDITIONS AS DESCRIBED BY WILSON AS WELL AS CERTAIN PECULIARITIES OF ASSOCIATED MOVEMENTS

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The case which serves as the basis of this discussion has been presented principally on account of two features, probably related. One is the *postural peculiarity of the left upper extremity* and the other is the *inability to perform certain movements of portions of this limb as isolated movements* and only as associated with other movements, especially with flexion of the forearm.

The former of these phenomena has recently acquired an added significance in connection with the article by S. A. Kinnier Wilson: "On Decerebrate Rigidity in Man and the Occurrence of Tonic Fits" (*Brain,* Vol. xliii, Part III, 1920). In this article attention is called to "certain analogies between the results of experimental mesencephalic transection in apes and other mammals and occasional clinical phenomena in man, in cases where cortical function is completely in abeyance." The type of cases is briefly "one in which there is evidence of withdrawal of cortical control in the form of unconsciousness or semi-consciousness, the result commonly of cerebral hemorrhage—submeningeal or intraventricular, or both—or of meningeal inflammation, or from the effect of certain intracranial tumors; also, for that matter, of hysteria. Coupled with this impairment of consciousness has been the appearance of tonic rigidity of trunk and limbs, minutely resembling experimental decerebrate rigidity, to which frequent exacerbations in the form of tonic or postural fits, accentuating and exaggerating the background of tonic posture, are superadded." Also "in certain types of organic and functional nervous disease these postural attitudes may be adopted involuntarily with full consciousness, and partial or fragmentary types also occur. To this last statement much importance is attached since the hypothesis is that the involuntary postures of a limb or segment of a limb in many forms of nervous disease, as well as the transient positions assumed in involuntary movements such as chorea or athetosis, are in reality simply parts of the complete decerebrate attitude."

Wilson accordingly considers his material under two main heads: I. "Those cases where the phenomena appear in association with loss of consciousness" and comprising "(1) Cases of decerebrate rigidity or attitude and tonic fits combined. (2) Cases of decerebrate attitude without tonic fits. (3) Cases of tonic fits without persisting decerebrate attitude." Group II includes: "(1) Cases of decerebrate attitude in conscious life. (2) Incomplete, limited, or fragmentary cases. (3) Cases of decerebrate attitude in the course of conscious involuntary movement, especially chorea and athetosis."

The salient features of decerebrate rigidity, as contrasted with the state following on transection below or in the lower half of the medulla are given by Sherrington as follows (*Integrative Action of the Nervous System*):

"Decerebrate rigidity is a condition which ensues on removal of the forebrain by transection at any of the various levels in the mesencephalon or at the thalamencephalon in its hinder part.

"If in a monkey or cat transection below or in the lower half of the bulb has been performed, the animal when suspended, artificial respiration if necessary being kept up, hangs from the suspension points with deeply drooped neck, deeply drooped tail, and its pendant limbs flaccid and slightly flexed. The fore limb is slightly flexed at shoulder, at elbow and very slightly at wrist. The hind limb is slightly flexed at hip, at knee and at ankle. On giving the hand or foot a push forward and then releasing it, the limb swings back into and somewhat beyond the position of its equilibrium under gravity, and it oscillates a few times backward and forward before finally settling down to its original position.

"To this condition of flaccid paralysis supervening upon transection in the lower half of the bulb the condition ensuing on removal of the cerebral hemispheres offers a great contrast. In the latter case the animal, on being suspended just as after the former operation, hangs with its fore limbs thrust backward, with retraction at shoulder joint, straightened elbow, and some flexion at wrist. The hand of the monkey is turned with its palmar face somewhat inward. The hind limbs are similarly straightened and thrust backward; the hip is extended, the knee very stiffly extended, and the ankle somewhat extended. The tail in spite of its own weight—and it is quite heavy in some species of monkey—is kept either straight and horizontal or often stiffly curved upward. There is a little opisthotonis of the lumbo-sacral vertebral region. The head is kept lifted against gravity and the chin is tilted upward under the retraction and backward rotation of the skull on the neck. The mouth is kept closed and there is some stiffness in the elevation of the jaw. When the limbs, or tail, or head or jaw are pushed from the pose they have assumed considerable resistance to the movement is felt, and unlike the condition after bulbar section, on being released they spring back at once to their former position and remain there for a time more rigid than before.

"The rigidity is immediately due to prolonged spasm of certain groups of voluntary muscles. The chief of these are the retractor muscles of the head and neck, the elevation of the jaw and tail, and the extension muscles of the elbow and knee, and shoulder (i.e., deltoids) and hip. In the dog and cat, just as spinal shock is more severe in the fore limbs than in the hind, so decerebrate rigidity is more marked in the fore than in the hind limbs. There is no obvious tremor in the earlier hours of its continuance; later it does sometimes become tremulant."

Wilson calls especial attention to the posture of the forelimbs in extension-pronation, and to the head retraction and opisthotonos.

Decerebrate rigidity combined with fits is illustrated by five cases, all of which showed during the seizures the above phenomena excepting the head retraction and opisthotonos which were not present in all. The forelimbs were also usually adducted. All were unconscious during the seizures. Two of the cases showed intraventricular and submeningeal hemorrhage, the latter extending to the cord, one due to a frontal lobe glioma; one case showed a glioma of the mesencephalon invading the upper part of the pons; a fourth case showed "a general meningitis over the cortex and more marked along the base. Section revealed an abscess in the right temporo-sphenoidal lobe, whence apparently the ventricular system had become infected. There was but slight internal hydrocephalus." The fifth case showed a "typical tuberculous meningitis of the base with moderate internal hydrocephalus." Interesting positions showing an incomplete decerebrate attitude were often present during the intervals between seizures. "The onset of tonic fits merely accentuated the already existing posture, showing that tonic fits are in reality attacks of decerebration, if the expression be allowed. The absence of any clonic element is of much significance. It may be taken to indicate that tonic and clonic movements have different origins and are the expression of the activity of different motor mechanisms; one is characterized by phasic and the other by static activity." Wilson points out that "the morbid process was so situated as to dissociate the cortex functionally from the lower centers"—shown also by the loss of consciousness—and that respiratory disturbances were also present.

Wilson apparently emphasizes the position of the upper extremities in the above cases on account of their presence partially or entirely in the fragmentary cases mentioned later. This, it seems to me, has led to insufficient emphasis on the fact that in all five cases the lower extremities were adducted, legs extended and feet inverted and plantarflexed. Attention is also called, by Wilson, to the fact that the position of the upper extremity (adduction, extension and pronation of forearm, flexion of hand and fingers) is the same as the typical hemiplegic position excepting that in the latter there is flexion instead of extension of the forearm, and also to the fact that "not a few cases of Little's disease conform strictly to the decerebrate type."

Decerebrate attitude without tonic fits is illustrated by one case, a child of fifteen months, unconscious with the above positions of upper and lower extremities continuously maintained for nine days. "The abdominals could not be elicited, the nerve-jerks were active and the plantars extensor."

Tonic fits without persisting decerebrate rigidity are illustrated, according to Wilson, by the so-called "cerebellar fits" and he further remarks: "It is, however, desirable, not merely to bring the phenomena of tonic fits into line with what experimental physiology has taught us of decerebrate rigidity, but also to show that *it is possible to resolve these complete seizures into component elements, that partial, incomplete, even segmental tonic attacks may be met with, and that some at least of the more or less fixed attitudes of head and neck, trunk, and of one or other limb or part of a limb, encountered in certain basal, subtentorial and other lesions, are in reality functional fragments, so to say, of a non-cortical motor mechanism which finds its full expression in decerebrate rigidity and in complete tonic fits.*"

"Tetanus-like seizures," whether of organic or hysterical origin, illustrate tonic fits without decerebrate rigidity and a case is given in detail where the above attitudes were shown and in addition a tendency to rotation. The fits were associated with respiratory disturbances and were followed by a completely atonic, flabby condition of the limbs. Another case exhibiting the decerebrate attitude, with some variation, had a glioma and intraventricular hemorrhage after operative interference. Other cases from the literature are cited, one of which is a case in which a tumor of the midbrain produced complete physiological decerebration at the level of the corpora quadrigemina.

One comment which may be made concerning the tonic fits especially is that in their case, at least, the exciting cause may not be so much the proprioceptive stimuli, mentioned later as maintaining the constant decerebrate attitude, as the irritative effect of the hemorrhages and meningitis.

An observation of Wilson's of especial interest, is the following: "The occurrence of tonic fits as a hysterical manifestation is a neurological platitude, but their real significance for the purposes of this paper consists less in their psychogenic origin than in the morphological aspect they may assume. If we imagine the most superficial degree of hysterical dissociation to involve the transcortical neuronic level the movements released will be purposeful and complex, resembling normal motor activity except for full conscious participation therein; if, similarly, we assume functional disintegration to descend to lower neuronic levels we should expect to observe phenomena morphologically analogous to those of decerebrate rigidity, which is, as a fact, precisely the case. In this way the actual movements of certain moderately severe hysterical seizures receive a physiological explanation and illustrate the disorders of function which we have associated with organic decerebrate human preparations. It seems unnecessary to cite any cases in support of this contention, so commonly are the opisthotonic and extension-pronation postures found in hysteria."

Wilson does not call attention to any relation between decerebrate rigidity and the picture of epilepsy, especially in its tonic phases, probably reserving this subject for later discussion. As is well known, there has been difference of opinion as to whether the tonic phase of epileptic seizures is due to cortical or subcortical mechanisms. If it be conceded that Wilson is correct in his comparison of the above clinical phenomena with those shown in experimental decerebrate rigidity and also in his localization of this tonic mechanism in the tegmentum of the midbrain, it is obvious that comparison with epileptic attitudes should yield valuable localizing indications whether or not the latter correspond to decerebrate attitudes.

Decerebrate attitude in conscious life is naturally apt to be not so complete or so indefinitely maintained but appears temporarily at least when the control of the corticospinal system is functionally or organically defective, there being "a conflict between two motor mechanisms, each attempting to jockey the other out of possession of the final common path." Two cases of infantile diplegia are given in detail illustrating this group.

Incomplete, limited, or fragmentary decerebrate attitudes form in some respects the most interesting group. One interesting case is of a child of two years in whom the left side showed the decerebrate extensor rigidity of both limbs while the right showed flexion at elbow and knee and abduction of the thigh, the head being turned to the right and tilted backward. Wilson points out that this case shows the type of rigidity seen in ablation of the left cerebral hemisphere, the extensor rigidity being homonymous. In this case "all the deep reflexes were active; the abdominals were present and the plantars of the flexor variety." The "cerebellar" attitude of the head (head inclined to one side and somewhat backward but turned to the other side) is cited as an example of a unilateral decerebrate head attitude. Wilson explains the opisthotonic attitude by pointing out that in bilateral cases the lateral head inclining and turning influences would balance each other but the head retracting influences on each side would reenforce each other, thus producing the opisthotonos. He points out that the above "cerebellar" attitude is not necessarily due to cerebellar lesions.

Passing to decerebrate rigidities limited to one limb, one segment of a limb or even "one phase of segmental movement," Wilson directs particular attention to the "pronator sign," which "is an important part of the complete decerebrate posture." Wilson shows the presence of this in one arm in cases of cerebellar lesions (homonymous side of middle lobe), of unilateral (homonymous) Sydenham's chorea (hemichorea), of diplegia with athetosis, of infantile cerebral hemiplegia with athetosis, of slight left hemiplegia with athetosis (following diphtheria). Another instance of limited decerebrate rigidity or attitude is the wrist flexion seen in hemiplegia, in the choreic hand (wrist flexion and some over-extension at the knuckles) and in the similar athetoid hand.

Wilson also mentions a case of a child (microcephaly with diplegia and athetoid attitudes) "whose physiological condition was one not far from complete decerebration, and in whom the vestibular reflexes of Magnus and DeKleijn were readily obtainable," i. e., extension of a previously flexed upper limb was elicited by passive lateral movement of head.

Decerebrate attitudes also occur, Wilson points out, in torsion-spasm (dystonia musculorum deformans).

The present case obviously is an excellent example of a partial decerebrate attitude involving especially the left upper extremity which shows the pronation phenomenon and the athetoid hand. It may be objected that the pronation at least is simply in such cases a result of the supination weakness so common in hemiplegia. That the two may be related is obvious but it is the explanation of the causes actively producing the spasm when certain cortical control is diminished which is sought and provided for in part by Wilson's point of view. It is doubtful too, whether there is any such supination weakness in chorea. The tendency to pronate in picking up and setting down small objects seen in some choreics may be another manifestation of the pronation phenomenon.

Another phenomenon, however, which does not invalidate Wilson's view but enters as a complicating factor into attempts at further analysis and explanation of abnormal involuntary contractions is the presence in hemiplegia and other pyramidal lesions of *flaccid* paralysis which, as is well known, is usually present for a variable period after the onset, may persist indefinitely or may even appear after a spastic condition has been maintained for a considerable period. Head has recently called attention to this latter phenomenon in his work on sensory disturbances due to cortical lesions. The writer has already expressed the opinion, not as a novel one however, that there must be other efferent cerebral paths concerned in tonus and that the pallio-ponto-cerebellar connections must be especially taken into account. According to some observers at least, it is the muscles which are not completely paralyzed or in which a partial recovery has taken place which show the spasticity. In this case an important difference would appear to be present between complete and incomplete pyramidal lesions.

THE MECHANISM OF DECEREBRATE RIGIDITY

Having considered the similarities between experimental decerebrate rigidity and certain clinical pathological attitudes, Wilson proceeds to discuss the mechanism producing this condition—the pathogenesis of decerebrate rigidity. This part of the article is also of great value, for sufficient attention is not usually paid to the actual nervous activities producing these phenomena.

Wilson first makes the point that the phenomena in question are not irritative but are "release" activities. Secondly he points out that the experimental work already quoted shows that for the development of decerebrate rigidity we are limited to the mesencephalon, cerebellum and pons (and upper bulb) inasmuch as the rigidity appears in transection above the midbrain and disappears in transection below or in the lower half of the bulb. It is admitted that decerebrate rigidity may also appear "when decerebration is higher leaving intact the basal ganglia, in particular the optic thalamus, and that movements analogous to those of tonic fits can be produced by stimulation of the latter structure."

Following Wilson's general position but not strictly his order of treatment, the next point is regarding the influences which maintain decerebrate rigidity. In the first place, the position of the animal is of importance. It is in the suspended animal where gravity is operative that it is most apparent. The rigidity thus developed is abolished (Sherrington) by deep anesthesia, after which it returns; it is abolished in a given part by cutting the posterior roots of that part, nor does it develop in that part if the posterior roots have been cut prior to decerebration; it is abolished in the hind limb by section of the ipsilateral column of the cord and in both hind and fore limbs by section of the ipsilateral ventrolateral column of the cervical cord. Section of the dorsal columns does not abolish it. Ablation of the cerebellum does not abolish it (Sherrington). "Bisection of or injury to the vermis, median section of the cerebellum and partial ablation of the cerebellum are all apt to produce the phenomena under discussion, but Sherrington is not convinced that the two conditions are identical, and especially states that after these cerebellar operations decerebrate rigidity 'sets in often, *but not always*' [italics mine]." (Wilson.)

According to Weed removal of the lateral lobe of the cerebellum diminishes the decerebrate rigidity in the ipsilateral limbs; removal of one half the cerebellum abolishes the rigidity in the ipsilateral limbs, but if removed before decerebration extensor rigidity is only slightly diminished on the same side; removal of the vermis before decerebration does not affect the development of bilateral extensor rigidity on removal of the cerebral hemispheres. He found, however, as have Sherrington and others, that stimulation of the anterior (superior) surface of the cerebellum, superior vermis and outward, abolishes the exterior tonus. Removal of the anterior portion of the superior vermis, according to Weed, is followed by the development of extreme extensor rigidity. Removal of the whole cerebellum, after decerebration and the development of decerebrate rigidity and without injury to the peripheral vestibular path and its nuclei, cause disappearance of the rigidity. Section of both restiform bodies and of the middle cerebellar peduncles has no effect; section of both superior peduncles results in marked initial accentuation of rigidity followed by a rapid and complete loss of all the rigidity. Section of one superior peduncle results in initial augmentation of rigidity on the same side followed by a marked diminution but never a complete loss; section of the three peduncles on one side and the inferior, middle and part of the superior on the other side produces a marked initial accentuation of the existing rigidity followed quickly by a complete loss of all rigidity on the side where all the peduncles were cut and nearly complete loss on the other. Median section through the mesencephalon produces an immediate accentuation and later a marked diminution with persistence of a slight degree of rigidity (incomplete decussation of brachia?). Application of warmth to the midbrain increases and cold diminishes the rigidity. Unipolar stimulation of the region of the nucleus ruber causes an accentuation of the contralateral rigidity at the elbow, a stronger current, relaxation of the ipsilateral elbow, still stronger the latter may become a flexor rigidity, stronger to both sides causes bilateral extensor rigidity. Stimulation of the colliculi has no effect. Transection just caudad to the caudal border of the inferior colliculi produces a short accentuation of rigidity followed quickly by a total loss of all rigidity and assumption of flaccid flexor attitudes. Weed also stimulated the position occupied by the frontal pallio-pontile tract from internal capsule to pons, in an animal with the hemispheres removed, and found the stimulation produced a complete disappearance of the ipsilateral and diminution of the contralateral rigidity. This was purely ipsilateral when a plate was interposed to prevent stimulation of the contiguous tract of the other side. This effect was abolished when the middle peduncle of the same side was cut. The frontal pallio-ponto-cerebellar path would thus be one inhibiting decerebrate rigidity. The inhibitory effect was diminished but not abolished by removal of the cortex of the anterior superior vermis. Weed concludes that the cerebellar cortex possesses some definite relationship to decerebrate rigidity but the function is double —an excitatory and inhibitory link both existing; that the main reflex center and, in that sense, the source of decerebrate rigidity is in the midbrain, probably the nucleus ruber; that the cerebellum is an important, perhaps absolutely essential path for impulses from limbs and trunk concerned in decerebrate rigidity; that the cerebellum forms an essential link in the inhibitory path from the cerebral cortex and that localization in the cerebellum, as far as can be ascertained from rigidities in the various extremities, are not sharply circumscribed or very minutely differentiated. From the fact that cutting the restiform body does not affect the decerebrate rigidity, Weed believes the path in the cord is the ventral spinocerebellar tract, which conclusion is apparently supported by the observation mentioned above as to the effect of cutting the ventro-lateral column of the cord.

Other important work bearing upon this rigidity-producing mechanism is that of Graham Brown who stimulated certain points in the cross-section of the midbrain at the level of the anterior colliculi in monkeys. These points corresponded to the red nucleus (and posterior longitudinal fasciculus). "Stimulation applied to this area of the cross section of the midbrain on one side evokes a state of flexion in the arm of the same side and a state of extension in the arm of the opposite side. On cessation of stimulation these reactions are continued as maintained postures. The movements of the arms are accompanied by movements of other parts of the body. The tail seems to bend to the same side; the lower limb of the same side extends while that of the opposite side flexes; the head is rotated in such a manner that the face is turned away from the side stimulated." These results can be obtained from stimulation of the midbrain after complete removal of the cerebellum and also after cutting the posterior roots of the parts involved.

In the experiments of Brown the animal was supported completely in all its parts and the tonic character of the response was shown by the "after-discharge." The fact that they could be elicited after cutting the posterior roots and after removal of the cerebellum simply shows that the efferent part of the mechanism was not affected by these procedures and that the latter would produce characteristic effects when stimulated artificially instead of being stimulated continuously by impulses passing upward by the usual afferent path or paths. Wilson recognizes this though in one place he seems to think that the possibility of eliciting reactions with the cerebellum removed invalidates the influence of the cerebellum in the maintenance of decerebrate rigidity. The maintenance of posture as well as the production of movement requires afferent as well as efferent impulses and it is difficult to see where the midbrain posture-maintaining centers receive their impulses except via the superior peduncle in the decerebrate animal, especially in view of the apparent absence of any collicular influence. About the only other path would be a path consisting of a series of short and possibly a few long ascending neurones in the reticular formation. Such a primitive ascending path is possible but hardly adequate.

Wilson then suggests that "in the red nucleus, perhaps in the pars magnocellularis, are represented combinations of the ventral cell-groups of the spinal cord, linked up according to their flexor, extensor, pronator, supinator, etc., functions." He also points out that the mechanism in question is probably on its efferent side more extensive than merely the red nucleus and the rubro-spinal tract and that "it seems desirable to point out that the nucleus ruber is but a part of Edinger's nucleus motorius tegmenti, which consists of associated motor-cell groups scattered throughout the pons and mesencephalon to the level of the ventral optic thalamus, including Deiters' nucleus, and that the effect of stimulation is not likely to be limited to one descending rubral path, viz., the rubrospinal. "There is in fact doubt as to the path or paths by which rubral activities reach the cell groups of the ventral cornua; further, rubro-pontine and rubrobulbar connections must not be ignored." The researches of Magnus and de Kleijn on the effect of passive movements of the head and change in position in space on tonus of the limbs are mentioned. Tremor may also receive a partial explanation because "tremor differs from rigidity, not fundamentally, but in degree."

In brief, in the above tegmental nuclei we have a mechanism which maintains tonus of the skeletal muscles, the activity of these nuclei is maintained or altered by proprioceptive impulses reaching them via afferent roots, thence probably, in part at least, by the ventral spino-cerebellar tracts, cerebellum and superior cerebellar peduncle, the cerebellum being also under an inhibitory (and other?) influence from the pallium. The tegmental nuclei are also under the more direct influence of forebrain mechanisms, notably the corpus striatum. In decerebrate rigidity there is a removal of forebrain influences, i.e., at least of those acting directly on the nucleus ruber, etc., from the striatum, of those acting indirectly via the cerebellum (pallio-ponto-cerebellar) and of those acting probably on the lower motor mechanisms, i.e., the pyramids. The resulting attitudes must be regarded as a composite of various influences, principally proprioceptive, acting, mainly by way of an uninhibited cerebellum, upon the tegmental nuclei in question.

It is certainly to be hoped that Wilson will soon make good his promise to give us further and equally illuminating papers upon this subject.

CASE ILLUSTRATING SOME OF WILSON'S PHENOMENA OF DECEREBRATE RIGIDITY

The patient, G. M. (No. 4445, general admission No. 95189), was first admitted to the Vanderbilt Clinic Sept. 2, 1915. She was then four years of age (would be five, January 31, 1916) and is therefore at present (Feb. 1921) nine years of age.

HISTORY. *Present Illness.*—The complaint noted Sept. 2, 1915, was a palsy of the left half of the body, which began two and a half months before. "While playing one day she fell and was paralyzed on the left half of the body. This has since improved to the extent that she can grasp objects with the hand, can walk with foot drop, and but slight facial weakness." Also "three weeks after palsy there was a general breaking out of purpuric spots all over the body."

More complete details obtained recently are as follows: The onset was about June 13, 1915, and at about 9 or 10 a.m. The patient was staying with an older sister (then about eighteen) who was in the next room. The sister heard the patient fall, ran in and thought she had fainted. She picked the patient up and laid her on the bed. She said that it (presumably unconsciousness) lasted about ten minutes. The patient was not able to walk immediately afterwards. The mother saw her that evening. She thinks the face was drawn to one side but is not sure which. No stiffness of arm or leg was noticed, nor any convulsive movements. The arms hung down when the patient was carried. No incontinence was mentioned by the sister. There were no headaches or vomiting. There have been no subsequent attacks. No somnolence nor insomnia have been noted.

The rash has recently been described by the mother as appearing about a week after the attack. It began like prickly heat, the spots got larger and red or purplish. It lasted a couple of days and was present, apparently, on arms, legs and trunk (face?). No temperature apparently was taken.

The patient fell out of bed the night before but there does not appear to have been any evidence that she was injured.

Birth.—It was a full term delivery, labor was not unusually prolonged and no instruments were used. There was no cyanosis nor asphyxia. The baby appeared normal in every way.

Development.—The patient was breastfed. The first teeth are said to have appeared at about ten months. She began to walk at sixteen months. The mother is uncertain when she began to talk. She walked perfectly before the attack. She began school at six years. Her school record appears to have been not especially good. She is now in class 3-A.

Injuries, Diseases, etc.—There is no history of severe injuries to head or other parts. There have been no operations. There is no history of convulsions, loss of consciousness (other than above), paralysis (other than above), diplopia, difficulty in eating and swallowing or speech. No history of visual or auditory disturbances, of vertigo, pains, headache, or vomiting.

The patient had diphtheria at six years of age. She has nat had chickenpox, measles, meningitis, mumps, otitis, pertussis, pneumonia, rheumatism, scarlet fever or typhoid.

Habits and General Physiological Status. The patient sleeps well, her bowels are regular, she has no incontinence of urine, nocturnal enuresis or other disturbance of urination.

Family History.—The mother has been married twenty-seven years and has had eleven pregnancies including one miscarriage and three deaths. The first two pregnancies are a boy and girl of twenty-five and twenty-four years respectively; bright, healthy and never had convulsions. The third pregnancy was a miscarriage at three months. The doctor said there was "a little inflammation." The fourth, a boy, died at nine years of age of diphtheria. He was not bright from birth and had one convulsion at about four or five years of age. The fifth and sixth, boys of nineteen and eighteen years of age, are healthy, bright and never had convulsions. The seventh died at eight months of marasmus. The eighth and ninth are boys of thirteen and eleven years of age and are healthy, bright and never had convulsions. The tenth is the patient. The eleventh died of meningitis after pneumonia. Was only sick three days. The father and mother are living and well. There is no similar trouble known in the family.

VOLUNTARY MOTOR SYSTEM. *Abnormal Attitudes and Deformities.*—When the arms are extended upward the left arm tends to assume an abnormal position which consists of pronation of the forearm (palm turns outward) and slight flexion (at times hyperextension), flexion at the wrist, hyperextension of fingers with thumb slightly adducted and hyperextended at the distal joint (Fig. 1). This attitude is present, less marked, when the arm is in the usual position while standing and is also elicited by various acts performed by other parts especially those involving attention and tension, e.g., when walking slowly on a line she tends to elevate, pronate, etc., the left arm; when she extends the left leg and resists its passive flexion, the left arm is often elevated above the head, pronated, etc. When running and at other times she at times carries the left forearm flexed, the hand dorsally flexed (overextended) and somewhat pronated with the fingers flexed (i.e., more nearly the hemiplegic attitude).

Gait.—A left foot drop is the only note on this in 1915. There is a slight left limp and perhaps a slight tendency to drop foot. Evidently the gait has markedly improved since the time immediately following the attack. Walking on the toes, on a line, backwards, sidewise and running are all as well done as ordinary walking, but walking on the heels is not done so well on the left, i.e., the toes of the left foot are not lifted so well from the floor. Fournier's tests did not show any new phenomenon.

Coordination.—The patient was formerly left-handed. She is now right-handed.

Equilibratory. Standing on both feet with eyes open and closed shows no ataxia. She

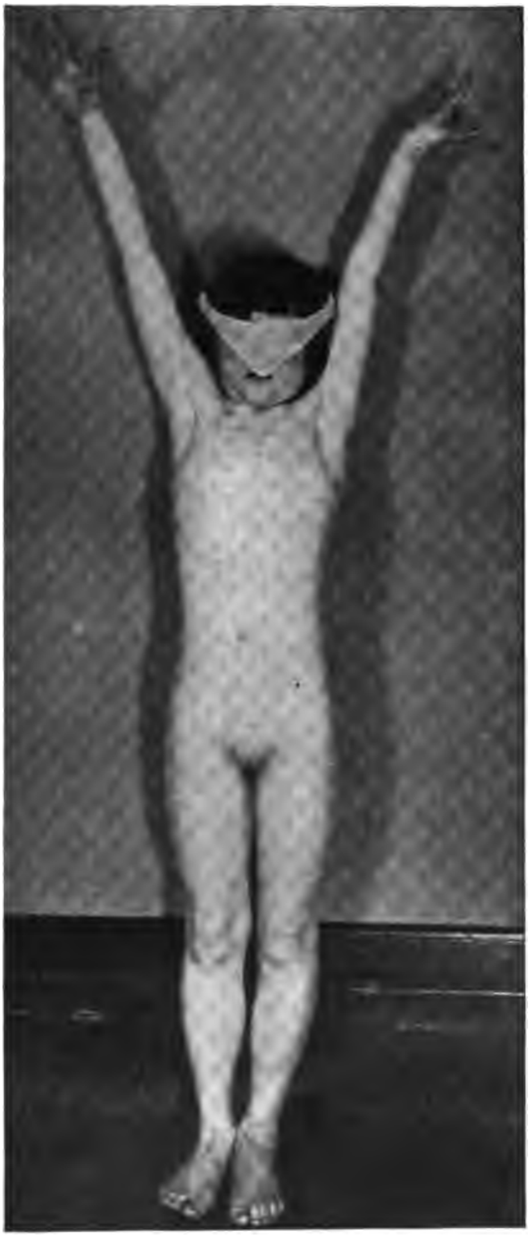

Fig. 1. This shows the hyperpronation of left forearm, the hyperextension of the left fingers together with flexion at the wrist ("athetoid" hand). There is also hyperextension at the elbow.

can stand much better on the right foot than on the left.

Non-equilibratory. The finger to nose test is performed without ataxia but when done with the left, this extremity assumes various abnormal attitudes during the act.

The patient can put each finger to the thumb in succession well with the right but not at all with the left.

The heel-to-knee test is performed well on both sides.

Skilled Acts.—Speech is normal in conversation.

Abnormal Involuntary Movements.—In the report of 1915 tremor was marked as absent There is none now excepting the associated movement mentioned elsewhere. When the patient was seen in 1915 and 1916 she was regarded, I believe, as showing some athetosis. These excessive movements have apparently disappeared or are represented simply by the more stereotyped associated movements or, rather, spasms evoked by certain movements and tensions.

Reflexes.—Deep: In the examination of Sept. 2, 1915, the biceps and patellar are noted as being greater on the left, there being no indication as to whether they are very active or slight. At present all of the deep reflexes are rather slight, the biceps being hard to elicit. They are somewhat more active in the left. There is no crossed patello-abductor, no tibio-abductor (periosteal) uncrossed or crossed and no ankle clonus. The left patellar, when reenforced, yields a rather spastic reflex.

Superficial: The abdominals all are active or very active and possibly somewhat greater on the right. There is a marked Babinski with fanning on the left which is easily elicited as a part of a general reaction to tickling, etc. There is a Chaddock and Oppenheim and a doubtful Gordon and Schäfer.

Muscle Strength.—The points noted in 1915 are: "Partial palsy of left arm, supination, pronation and extension of fingers especially weak." There are at present the following pareses: Slight pareses of the left shoulder (elevation) and of the left arm, apparently all movements perhaps greatest in rotation inward. The left forearm shows a slight weakness of flexion, extension and pronation and a marked weakness in supination. The left hand shows an interesting condition. Its various movements (flexion, extension, abduction and adduction) cannot be performed at all unless the forearm is at the same time flexed. When this is allowed, the strength is normal or nearly so. The same applies to flexion and extension of the fingers except there is here a considerable diminution in strength. There is still greater diminution of strength and limitation of movement in abduction and adduction of the left fingers.

The left thigh shows slight pareses of adduction, abduction, rotation in and rotation out (most marked in the last) and possibly of flexion and extension. The left leg shows some weakness of flexion. The left foot shows a marked weakness and limitation of movement of dorsal flexion and also a weakness, less marked, of abduction. The left toes show marked weakness and limitation of movement in both extension and flexion. The *voluntary* extension of the left big toe is weak but when reenforced by plantar or other stimulation (passive flexion of left leg) or elicited by the latter alone it is of more nearly normal strength.

Muscle Status.—No marked atrophies were noted. The left hand and forearm show some general atrophy. The various parts of the left upper extremity, especially the fingers and hand, are more extensible than the right. There appears to be possibly also a slight hypotonus elsewhere, including the lower extremities.

Abnormal Associated Movements.—The pronation sign of Strümpell appears to be absent, also the thumb flexion sign of Hoffmann. The patient cannot synchronously adduct the thumb and all the fingers, either the thumb or little finger remain partly abducted. She cannot resist passive extension of her flexed fingers (excepting the thumb which she is told to keep extended and abducted) without flexing the thumb. This is true of both sides but is more marked on the left. Passive hyperextension of the extended fingers also produces flexion of the thumb. Associated fist-clenching is not present. The trunk-thigh sign of Babinski appears to be absent. She is able, when recumbent, to elevate both lower extremities with legs extended (Grasset-Bychowski). The tibialis sign of Strümpell is absent The coughing sign (leg extension on coughing) is present on the left. The reenforcement sign is absent.

When the patient is recumbent, strong extension of the left leg (e.g., holding it in the extended position against strong opposition) elicits extension of the left forearm and fingers with flexion at the wrist and even elevation of the arm. On the other hand, strong flexion of the leg at times elicits flexion of the forearm. Extension and flexion of the right leg does not appear to have

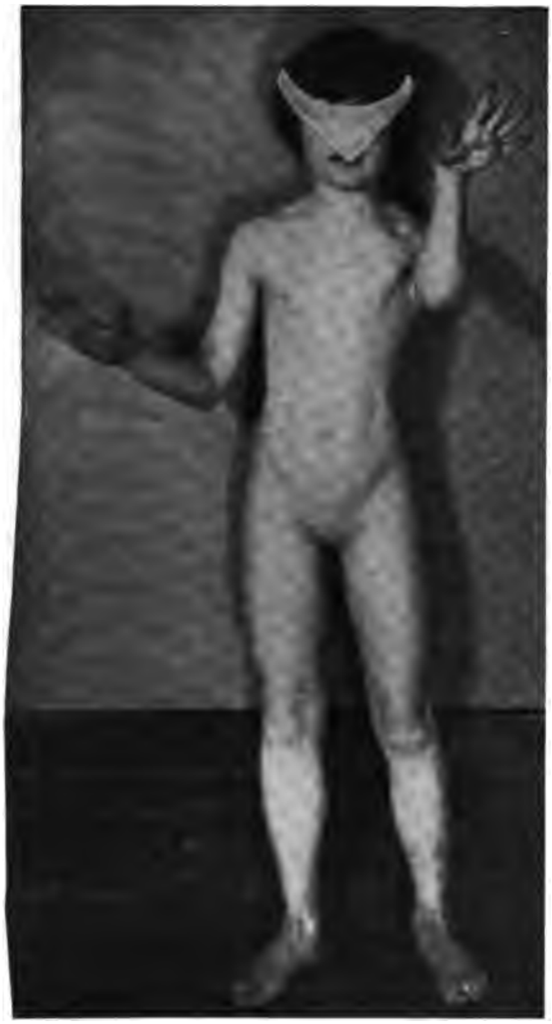

Fig. 2. This shows the patient at the instant of having accomplished the opening of her hand by associated flexion of the forearm. The right upper extremity also shows a contralateral associated movement.

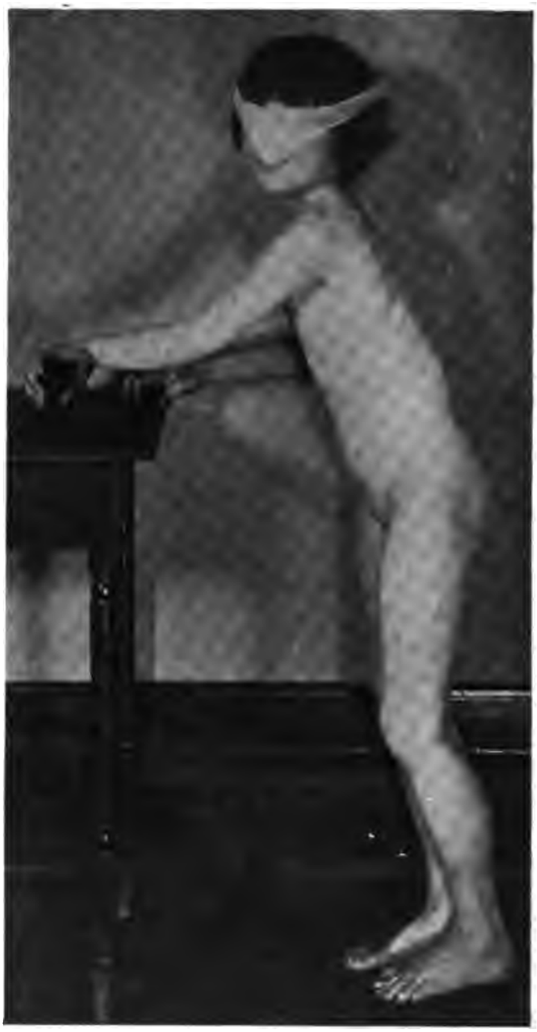

Fig. 3. This shows the patient attempting to release the object in her left hand. The pronation of the forearm is shown. The intensity of her effort is not brought out by the photograph.

this effect upon the left arm, certainly not to the same extent.

Passive movements of the head, against resistance, do not appear to have a definite and constant effect upon the movements of the left upper extremity. Both this point and the preceding require still further study.

Changes in position of the head without movement did not appear to have any marked effect.

Perhaps the most interesting phenomenon is the inability of the patient to close and especially to open the fist or to release grasped objects quickly. This she usually accomplishes by making associated movements, especially flexion of the forearm, at the end of which the fingers open quickly (Fig. 2). This inability is consequently much increased by holding the forearm in extension and preventing its flexion, thereby preventing the opening of the fist as an associated movement. Holding other parts may have some influence but this is partly due to emotional effect perhaps. The inability to release is enormously increased by emotion. In taking a kymographic record of these movements, the patient was alarmed because she thought electricity was to be used and was then at times almost indefinitely unable to release an object grasped with the left hand. It is also interesting to note that in trying to release (and also often in grasping an object) she excessively pronated the forearm as a severely affected choreic often does (Fig. 3). Diversion of attention also helps her in opening the hand.

General Sensory.—In the report of 1915, "sensation" is indicated as normal. There are now no disturbances of touch, pain, temperature (cool and warm), vibratory and muscle-tendon sensibility (arms, hands, fingers, legs, feet and toes being tested for the last named). At times muscle-tendon sensibility (sense of position and movement) appeared to be not quite as good in the left fingers as in the right but this was apparently due to inattention and possibily an indirect effect of the different tonus of the left hand.

Cranial Nerves. *Optic Apparatus.*—Vision was not tested. The fields, roughly tested, are normal. The fundi are normal. The pupils are normal, equal and react well to light and convergence. No abnormalities of position or movements of the eyes are present. There is no nystagmus, only a few nystagmoid movements are elicited when the eyes are directed laterally. The palpebral fissures are equal. There is no ptosis and no von Graefe.

Acoustic.—This was not carefully examined but hearing appears to be normal on both sides.

Trigeminus.—The corneal reflexes are active.

Facial.—The record of 1915 states that there is a left facial weakness. There is at present a very slight facial weakness apparently principally or entirely of the lower face. It is interesting to note that the patient can wink the right eye better than the left.

Glossopharyngeus and Vagus.—The movements of the palate during phonation are normal.

Hypoglossus.—The tongue curves slightly to the right when protruded but there is probably no abnormality here. The various movements of the tongue are performed well. It shows no atrophy.

Mental Status.—The impression is that intelligence, emotion, attention and memory are normal.

Systemic.—No tegumentary abnormalities were noted. The heart is normal. In the report of 1915 it is noted that "blood pressure is 95."

Laboratory Tests.—The report of 1915 states that the blood Wassermann was negative. An *x*-ray taken Dec. 1, 1915, of the right side of the head is stated (Dec. 8, 1915) to show nothing.

THE MECHANISM AND SIGNIFICANCE OF ASSOCIATED MOVEMENTS

The phenomena included under the term "associated movements" have close relationships with other allied phenomena such as various "involuntary movements," movements due to emotional states and infantile movements, and the distinction between them cannot always be made. In the first place it is obvious that all "acts" are associated movements. By the term associated movements, however, is usually meant, I take it, those movements which are made during the performance of an act or movement but are not an integral or necessary part of it, movements which are elicited by an active or passive movement or movements of some other part but not mechanically necessitated by the latter (i.e., not inevitable on account of mechanical arrangements of tendons, bones, etc.). In the normal condition such movements are often obviously initiated by the primary voluntary movement or act and they can be voluntarily suppressed but they appear without *direct* voluntary initiation and, in fact, may require voluntary effort for their repression. They can, however, also be reproduced voluntarily.

From this latter point of view, one test of voluntary control is the ability to suppress such accessory or associated movements either as a whole or to break up the associated movements into their component parts. This is only putting in a somewhat different way the ordinary view that one salient feature of what we call voluntary control is the ability to perform isolated movements. This ability is thus one of the tests of voluntary control as is shown in such tests as successively touching each finger to the thumb of the same hand and performing other single finger movements. Opposed to such movements we have the simultaneous finger movements whether of flexion, extension, abduction and adduction which we all know to be more easily performed than the above movements of individual fingers, and the above peculiarity as well as the, so to speak, involuntary elicitation of associated by voluntary movements has led to their designation or qualification as "automatic" associated movements. The formation of the skilled act or acquired group of associated movements from this point of view consists in the substitution of a group of movements more or less different from the automatic associated movements. The piano player when playing chords, especially octaves, may have the fingers set and moving in much the automatic position but the arms move in a dissociated manner. In playing scales and in "fingering" in general the finger movements are dissociated. Certain modifications of the Klippel-Weil test also illustrate these points. As the writer understands it, the test itself consists in passively extending the involuntarily flexed fingers of the patient. The organic nature of the nervous affection causing this flexion is shown by the automatically elicited flexion of the thumb. Passive hyperextension of unflexed fingers is also performed as a test to elicit this reaction. The mechanism upon which this rests is further displayed, probably, in the following. Anyone can extend and adduct the fingers and at the same time abduct the thumb, nor is it difficult to flex the adducted fingers without flexing the thumb. If, however, the fingers of another be inserted in the clenched fist and passive extension be attempted, the person resisting this passive finger extension will feel a distinct tendency to flex the thumb. It is this normal tendency to associated thumb flexion which underlies, I take it, the exaggerated form seen in pathological cases and in the permanently flexed thumb of a hemiplegic contraction. The uneducated toes are usually only capable of simultaneous plantar flexion, of simultaneous extension, with abduction, or "fanning," and of extension of the big toe and flexion of the others, the latter being of course identical with a typical "Babinski." The bilateral (or contralaterally evoked) associated movements are also illustrated in normal conditions by the relative ease in executing identical movements with the paired extremities.

From the above it is obvious that there are really two categories of repression of automatic associated movements, one the repression of *accessory* associated movements, the other the breaking up of an "automatic" association and the substitution of a "learned" or skilled association.

Both the capacity for performing isolated movements and the tendency to associated movements (as seen in the above Klippel-Weil sign) should be studied systematically in normal as well as abnormal individuals. The performance of isolated movements is a test of motor control and is notably defective in young children and mental defectives as every one is aware when getting them to perform certain movements in testing their strength. Most persons can draw one side of the mouth to one side, elevate one side of the nose and wink one eye. Very few can elevate one eyebrow. The lateral hand movements as opposed to flexion and extension are the more difficult and harder to elicit in children and the same applies to lateral movements of the foot. Another example of the tendency to automatic association is shown by the pronation sign of Strümpell. The easiest position to flex the forearm is semipronation, i. e., so that the radial margin comes next the arm. Extension at the wrist is the natural accompaniment of clenching the fist and slight flexion at the wrist and abduction of the fingers is the natural accompaniment of abrupt and forcible extension of the fingers (athetoid position). It is obvious that in all such cases mechanical peripheral arrangements must also be carefully considered. It is very possible that peripheral arrangements are better adapted to the older types of movement than the newer which partly replace them and that the "voluntary" movements not only have partially to inhibit older central neural mechanisms but also to use peripheral arrangements which are not completely adapted to their performance. There are arrangements of tendons, for example, which certainly interfere with completely isolated finger movements. Whether we must also take into account more than one system of somatic effectors and peripheral nerves innervating them I will leave to Dr. J. Ramsay Hunt to answer.

It is also obvious that many of the associated movements are, as often elicited, of a spasmodic or tonic character, e.g., the Klippel-Weil.

It is not proposed at present to discuss the classification of the associated movements as done by Foix and others.

The distinction between the automatic associated movements and the abnormal involuntary movements is not always easily made. The main distinctions, I take it, are that the former are more definite or limited and that the latter are apparently more spontaneous, at least more easily elicited. As is well known athetoid movements can be elicited by passive opposed movements of other parts and choreiform movements are increased in the same way. In such cases they would appear to fall partly in the category of associated movements. In fact Lewandowsky has called athetosis a mass of associated movements. The spontaneity of such movements is more apparent than real. They usually disappear during sleep and also diminish or disappear when the part is completely supported, and other parts are at rest. The present case might be called by some an athetosis or an athetoid condition.

The close connection between associated movements and movements of affective expression or, rather, the overlapping of the two, is evident when we consider that many of the latter are associated movements (*vide* the expressionless face of the paralysis agitans when the facial associated movements are absent) and that they are often brought out by emotional states. The influence of the latter on the patient was very obvious, especially upon the spasm and inability to release her grasp. It is the accessory (shall we say the supernumerary?) associated movements which give the flavor, the pleasing character and animation often, to movement. On the whole, however, perhaps the affinity is greatest between the affective movements and abnormal involuntary movements. The case of tremor is obvious, but aside from that the grimacing of athetosis and chorea and the writhing and agitated movements of some affective disturbances as well as various hysterical movements may be cited, aside from the great influence of affect upon both.

The relation of associated movements and the abnormal involuntary movements to the infantile movements is both important and obvious. Strange to say the normal baby has been studied with some care by the psychologist and behaviorist but has been neglected by the neurologist. We probably all have thought of the infant as subject to choreoathetosis with a considerable admixture of convulsive movements but have not studied him intensively. We know he begins with a tendency to fist-clenching which later as in some hemiplegics may loosen up into an athetoid position at times when the "post-hemiplegic" condition is reached, with a better development of the pyramids and a better release of the striate system possibly. The athetoid position of the fingers is often seen in good illustrations showing childish poses.

Reference has already been made to the fact that many of the associated movements are more of the nature of associated spasms and it would seem that all categories of movement, not only the automatic associated movements and the volitional movements but also reflex movements have a tonic or postural counterpart. We know that the tonus or posture of each muscle is maintained reflexly by impulses from itself and that as shown by the work of Brown, Weed and others just cited there are automatic mechanisms for posture as well as movement, mechanisms which may explain certain hysterical phenomena as well as certain phenomena due to organic disease. We also know that certain postures, as well as certain movements represent the highest volitional mechanisms of the nervous system. This is, in part at least, the position recently advanced by J. Ramsay Hunt. We all recognize and often see statues and pictures embodying postures which clearly indicate cortical activities—in fact perhaps the majority of statues represent at least an admixture of cortical and emotional expression with the more purely automatic. To one who has watched wrestling it becomes quite a problem how far the contestants' activities are automatic, how far emotional and how far planned. The present champion, Lewis, relying on his great weight and strength, often frees himself from a dangerous hold by thrashing movements exactly similar to those of many lower forms in a similar predicament. They are emotional-automatic in character. On the other hand in seizing an opportunity for applying his famous and carefully developed headlock there is a much greater cortical element. No act nor pose, however, can be said to be purely of one character or another. Probably all acts, however skilled, require the automatic cooperation of fixation and of accessory movements.

It must also be remembered that the so-called automatic associated movements are under voluntary control—they can be voluntarily produced and disappear in complete voluntary paralysis.

Regarding posture in decerebrate rigidity and its relation to standing it should be pointed out that this should vary according to the normal posture of the animal. In fishes, or water animals generally, it is evident that no anti-gravity mechanism would be present similar to that in air animals or those depending upon a fixed or firm support. Again, in the latter the fundamental postures vary in different forms. Sherrington has pointed out that the rigidity assumed by the frog is one of flexion, i.e., the squatting attitude of the animal. Applying this to man it is evident that the upright position entails further changes. That the mechanism of standing and walking in many quadrupeds is of a fundamental or paleoëncephalic character is shown by the fact that they stand at birth and by the observations of Graham Brown on cat fetuses in which the progression type of movement could be demonstrated. However much this may be regarded as demonstrating that the acts of standing and walking are not "learned" in these forms, it hardly applies to the infant when he painfully changes from creeping to standing and walking. It is especially the assumption of the standing position that is new, i.e., that is "learned", for the alternating arm and leg movements seen in creeping are obviously from the above discussion in part an old mechanism. In fact the whole change to the upright position is closely correlated in more than one way with the development of the neëncephalic apparatus which reaches its culmination in man.

When we try to ascertain the neural mechanisms underlying these phenomena we soon realize how defective our knowledge is. This is especially the case when we try to locate exactlv the efferent integrative mechanisms at the various levels.

In a broad way, in the central nervous system there are on the efferent somatic side at least four integrative levels or rather four kinds of integrative levels, each possibly divisible into a number of categories. Beginning with the lowest we have the lower motor neurones upon which all impulses which change the position or tonus of the striated voluntary musculature are concentrated or integrated. This is the case whether or not we subdivide them into several categories. Secondly, there is the ground bundle-reticular formation of cord and brain. Thirdly, there is the older part of the suprasegmental mechanisms of Meyer comprising the colliculi, portions of the cerebellum (the paleocerebellum) and the older portions of the forebrain, on the efferent side mainly the globus pallidus and related structures. Fourthly, there is the major part of the efferent pallial apparatus and its adnexa such as the neocerebellum. The principal neopallial descending paths are the pyramidal, the pallio-ponto-cerebellar and the pallio-thalamic. The first of the above four are the efferent segmental or peripheral, the second the intersegmental and the third and fourth are the suprasegmental portions of the nervous system.

The reticular formation in a broad sense includes also the nucleus of Deiters and the nucleus ruber, the latter especially being practically differentiated out of it.

The cell groups of the reticular formation, as distinguished by the Nissl method, are, according to the systematic observations of Jacobsohn, as follows: (1) Cells of medium size and polygonal character which are found more or less throughout the whole formation. These he regards as "intercalated" neurones (Schaltneurone). (2) Small, rounded cells also diffuse in location and which he regards as sensory (sensibler). (3) Pyriform or spindle-shaped cells, pale and homogeneous in appearance. These cells occupy the region of the raphé of the upper bulb and are termed the nucleus pallidus raphés. (4) Very large giant cells—the largest cells in the medulla—of homogeneous appearance and therefore regarded as sensory. They lie in the upper level of the bulb and the lowest level of the pons. They are intermingled there with the motor reticular cells (see below) of this region and with the cells of the nucleus pallidus raphés. (5) Large cells resembling the cells of motor nuclei. These cells are scattered throughout the reticular formation. There are few in the caudal region of the medulla. They increase in number up to the caudal portion of the pontile tegmentum. There are fewer in the pontile tegmentum with some increase in the peduncular tegmentum. The large-celled portion of the nucleus ruber belongs to this group, also, probably, the interstitial nucleus of Cajal and what is sometimes known as the large-celled portion of the nucleus of the posterior commissure (not the nucleus of the posterior commissure *sensu stricto*). Probably some of the cells of the subthalamic and hypothalamic region should be included. Deiters' nucleus also belongs in this category. It may be added here that the cells of Betz, probably the large cells of the globus pallidus, certain cells in the colliculi and the cells of the nucleus fastigii of the cerebellum are of similar character.

Cajal divides the cells of the reticular formation into two types: (1) Small and medium sized cells and (2) large and even giant cells. The latter are spoken of as resembling motor cells and Cajal evidently does not distinguish the "pale" or "homogeneous" giant cells of Jacobsohn nor does he subdivide the smaller cells into the types mentioned by the latter author. The small cells do not stain well by the silver methods and Cajal's descriptions of the connections of the cells of the reticular formation are of the large cells. Very briefly stated, he finds that the axones of these cells by turning or branching become longitudinal fibers. They are both crossed and uncrossed. These longitudinal fibers lie principally on the outskirts of the reticular formation especially near the raphé, including the posterior longitudinal fasciculus, near the dorsal border of the gray reticular formation and also to some extent along its ventro-medial border next the pyramidal tract. While the fibers are both ascending and descending, they are predominatingly descending. They usually give off collaterals before becoming longitudinal. It is evident that this, in a general way, is the position occupied by the vestibulo-spinal tracts, and it is well known that in general reticulo-spinal (other than vestibulo-spinal and rubro-spinal) fibers occupy these positions but they are often overlooked in discussions involving these systems of fibers. Regarding the fibers which terminate around the large cells of the reticular formation, Cajal finds they are (a) collaterals from the longitudinal axones of the secondary sensory neurones; both those of bulbar nerves and also from the remnant of the ventro-lateral column representing ascending tracts from the cord, and (b) collaterals of the pyramidal tract. The former are more abundant. Cajal also finds the large cells enveloped by "terminal buttons" representing the terminations of the above collaterals. The small cells of the reticular formation which stain feebly in the reduced silver preparations do not show these pericellular terminations. Cajal concludes that the cells of the reticular formation are not all alike functionally, that the majority are sensory neurones of the third order receiving collaterals from the secondary sensory tracts and the minority are motor neurones of the second order, transmitting to the bulbar motor neurones the impulses they receive from the pyramidal tracts. The fibers in the gray and white reticular formation, according to him, arise from four principal sources (leaving out of account fibers from acoustic nuclei, cerebellum and colliculi), i. e., (a) ascending fibers from the cord, (b) ascending and descending fibers from the interstitial motor (second order) cells, (c) ascending and descending fibers from sensory cells of the third order, and (d) long and short sensory paths of the second order.

Two comments may be made on the above, first that the observations as to collaterals from the pyramidal tracts were made upon fetal or very young mammals and they are hardly conclusive as to the existence of such collaterals in older animals. Cajal has made similar observations as to collaterals from the pyramidal tracts terminating in the nuclei pontis and also of collaterals from descending pallial fibers entering the corpus striatum and other basal nuclei. The importance of these observations in trying to arrive at an understanding of the precise neurological mechanisms involved in the performance and acquisition of skilled movements in particular is very great and it is desirable that the presence or absence of such collaterals be definitely determined in the adult human brain. The Marchi method has its limitations in settling points of this nature and in general it may be added our knowledge of the exact termination of the pyramidal fibers is greatly lacking in precision.

A second comment upon Cajal's account is that the relations of the numerous smaller reticular formation cells are left out of account, for Cajal in speaking of sensory cells of the third order apparently is only speaking of the large cells.

It may be added that collaterals from secondary sensory paths also terminate directly in the lower motor nuclei of bulbar cranial nerves and, in the case of the trigeminus, collaterals from the mesencephalic root terminate in the motor nucleus of this nerve.

That reticulo-spinal fibers pass downward the whole length of the cord, at least as far as the vestibulo-spinal, interstitial nucleus and rubro-spinal contingents are concerned, is well known. In the cord they occupy a similar position to that in the medulla, allowing for differences due to the external addition in the medulla of the more recently enlarged inferior olivary nucleus. As is also well known, other reticulo-spinal fibers pass downward for at least a part of the length of the cord and it is possible that the tracts known as the vestibulo-spinal and the fibers from the interstitial nucleus include other reticulo-spinal fibers.

Besides receiving terminations from ascending cord fibers, superior peduncular fibers (to the red nucleus), "juxta-restiform" fibers (to vestibular nuclei) and lenticulo-rubral fibers, it should be pointed out that the other portions of the reticular formation also receive fibers from the superior cerebellar peduncle, and that the practical extension of this formation into thalamic regions opens a possibility for other connections not definitely known. Finally there is the work of von Monakow on the nucleus ruber according to which this nucleus has double (i.e., afferent and efferent) connections with the thalamus and with the prefrontal and centro-opercular regions of the pallial cortex, the afferent (to the ruber) being perhaps the more important, and also according to which the ruber sends crossed descending fibers to other parts of the reticular formation. The part of the nucleus ruber having the above connections is the larger, cephalic, smaller-celled portion of the nucleus which is the new portion, the older caudal, large-celled, portion, much smaller, giving rise to the crossed rubro-spinal tract. The most important afferent connection to the newer portion is the superior cerebellar peduncle.

In the cord there do not appear to be any cells similar to the large motor reticular cells unless they be certain large cells found in the lateral part of the intermediate gray. There are nevertheless, as shown by Sherrington in the dog by means of his "successive degeneration" method, long descending intersegmental fibers extending from upper thoracic to lumbar cord. They soon assume the external position characteristic of long tracts and are present in both lateral and ventral columns, near the ventral fissure especially. There are also long ascending intersegmental fibers. As already mentioned, however, it is doubtful whether these long intersegmental tracts of the cord can be regarded as analogues of the reticulo-spinal fibers from motor reticular cells.

The reticular formation may also receive some fibers from the cerebellum *via* the middle peduncle (perpendicular pontile fibers) some of which may terminate in the superior central nucleus of the reticular formation.

Among the ascending fibers of the reticular formation are some which probably terminate in the thalamic or subthalamic region dorsal to the field H of Forel.

The destinations of the axones of the smaller cells of the reticular formation are obviously less well known than those of the large cells. Certainly many of them are short intersegmental or intra-segmental fibers. It is possible that the numerous fibers from the reticular formation of the lower bulb which pass down in the ventro-lateral white columns to terminate in the cervical cord (bundle of Thomas) and which are smaller than the long descending reticulo-spinal fibers of the cord are from such smaller cells.

It is obvious that the reticular-ground bundle formation thus sketched is one consisting of long and short intersegmental neurones and mediating between the afferent and efferent fibers probably in the performance of various reflexes of varying complexity, that it is also under the influence of cerebellum, midbrain and forebrain and that it may thus also constitute manyrelayed ascending and descending forebrain paths. It is the oldest associative mechanism of the vertebrate central nervous system—the one necessary for vital neural activities and probably the most important one in forms without well developed cephalic sense organs and suprasegmental apparatus. It is the primitive higher, cephalic, motor apparatus.

The part of this apparatus which is most definitely known is the large-celled nuclei and it seems probable that their designation as motor is correct inasmuch as they form long, principally descending, tracts to motor root nuclei. That the posterior longitudinal fasciculus is the primitive somatic motor path has been pointed out by Johnston and others but probably the conception should be enlarged to include the other long descending reticular systems. Added to this, or lifted out of it, are the suprasegmental hind, mid and forebrain motor systems of the internal cerebellar nuclei, certain collicular cells and certain cells of the globus pallidus. Finally, and more independently, the neopallial pyramidal system is added. The former, certainly the cerebellar, and probably in part the pallidal, act only through the intervention of the motor reticular apparatus, i.e., they terminate in it; the pyramidal apparently, as shown later, acts largely or entirely independently of it, i.e., passes by it.

The exact terminations of the motor reticular formation fibers are not known excepting those from Deiters' nucleus to the oculomotor nuclei where actual pericellular terminations have been demonstrated in some forms. The position of the vestibulo-spinal and mesencephalo-spinal fibers in the cord make their termination directly around the ventral horn motor cells probable, though even here there are also present bodies of intersegmental or intrasegmental neurones. The position of the rubro-spinal tract makes the direct termination of its fibers around the ventral horn cells less probable. Another reason against their direct termination is the possibility that there are too few fibers to innervate the motor root neurones. When we consider, however, the spreading dendrites of the former and the presence (in young mammals at least) of collaterals from their axones and also the possibility of collaterals and widespread and terminal arborizations of the descending reticulo-spinal fibers, this reason does not appear conclusive. If the latter do not terminate directly around the motor root neurones we must, of course, infer that they terminate around the spinal intersegmental neurones, which in turn link the motor root neurones.

The question as to the exact termination of the pyramidal fibers is perhaps still more obscure. As is well known there is difference of opinion as to whether those of the lateral tract terminate in the intermediate gray or in the ventral horn. Careful investigations with the Marchi method (e.g., Lewandowsky's) have not shown the Marchi granules extending into the ventral horn or into the motor cranial nerve nuclei, but negative evidence with this method is not entirely decisive in a question of this kind. The position of the lateral tract is also against its fibers terminating in the ventral horn, for the usual general arrangement in the nervous system, in the cord at least, appears to be for terminals and collaterals to terminate in the gray lying next to the fibers in question. It would be interesting and might throw light upon the so-called inhibitory influence of the pyramids if it could be shown that they act upon the lower motor neurones in a different manner or by a different route than the motor reticular neurones. In this connection it may be recalled that the superficial reflexes unquestionably pass to the lower motor neurones *via* intermediate intersegmental neurones, while certain tonic or tonus-maintaining impulses and possibly those producing the deep reflexes not improbably pass directly *via* afferent root collaterals to the lower motor neurones. This and the antagonism between pyramidal influences and tonic reflexes on the one hand and the cooperation between pyramidal influence and superficial reflexes on the other would lead us to look upon the pyramids as acting upon intersegmental neurones, especially upon those receiving terminals of fibers of superficial sensation. On the other hand the large size of the pyramidal tracts and the precise and limited character of many of the reactions they perform, would lead us to favor the direct termination of their fibers on the motor root neurones, as in this way, apparently, the cortical motion combination would be best preserved. In the brainstem the neurones receiving terminals of superficial sensory nerves (secondary sensory neurones) do not receive pyramidal fibers.

When we further consider that the physiological significance of even the grouping of the lower motor neurones is not entirely clear and, still further, that possibly not all these neurones have the same tonic and kinetic significance it is evident we are still far from having that clear knowledge of the exact integrative arrangement in these lower levels which is necessary to give movement a clear neurological explanation, even aside from the question of the part played by the various higher mechanisms of cerebellum and forebrain.

The possibility of pyramidal collaterals has already been mentioned and the question arises whether the reticulo-spinal apparatus may not also be under pyramidal control. This would give an explanation for the associated movements seen in slight pyramidal lesions and account for the voluntary control of such movements in normal conditions. In view of the fact that pyramidal fibers can only be traced into the pontile tegmentum and not to definite cells, this possibility cannot be entirely excluded on purely anatomical grounds. If, however, this be the case, there should be a difference clinically between lesions of the pyramidal system above the midbrain, where the alleged fibers to the reticular formation begin to be given off, and below this level. As far as the writer is aware there are no such differences, excepting possibly the differences between paraplegia in flexion and in extension to which attention has been called by Walshe. It must be remembered, though, that in many regions injury to the pyramidal fibers is liable to affect tracts in its vicinity which may obscure the picture. In the pes, the pallio-pontile fibers are liable to be involved, in the pons both the pallio-pontile and ponto-cerebellar systems and in the cord the rubro-spinal tract—one of the very tracts it is desirable to exclude. Basal bulbar lesions such as those producing the hypoglossal type of alternating hemiplegia, without deep sensory involvement, would be the best to give a "pure" pyramidal picture. Above the midbrain, capsular lesions obviously may also involve not only pallio-pontile but other systems as well. Nevertheless a more intensive study of selected cases of hemiplegia and paraplegia, especially pursuing a combination clinical method such as that used by Head in his study of sensory paths in the cord, might yield results even without complete autopsy material.

Other forebrain neurone systems possibly or probably acting upon the reticular formation are strio-rubral, possibly other strio-reticular fibers (e.g., to the reticular neurones giving rise to the posterior longitudinal fasciculus) and pallio-rubral neurones.

The nature of the impulses conveyed by these neurone systems is certainly far from clear. It is not even clear whether they are tonic or kinetic or both. The fact that the removal of the forebrain systems results in increased tone, maintained, as already seen, by constant proprioceptive stimuli acting upon the reticular formation, largely via the cerebellum, does not prove either that the various forebrain impulses are simply inhibitory or that they are all of the same character. The performance of any definite act is usually attended by other movements not so essential (associated movements), by the "setting" of certain parts to afford a fixed base for the movement or movements ("fixation") and suppression or diminution of activities (including tonicities) antagonistic or even unessential to the performance of the act (inhibition). It thus hardly seems probable that any given neurone system is *solely* inhibitory in function though here perhaps we enter an obscure region in neuro-physiology. The pyramidal system is usually conceived as causing active contractions and not simply inhibiting a lower tonus-maintaining mechanism. Consequently we should look for movement as well as inhibition as part of the function of the above forebrain neurone systems. On the other hand the reactions to stimuli applied to the nucleus ruber, etc., by Graham Brown, while they were movements, were characterized by an after-discharge and were thus tonic in character. They were also different from those elicited by stimuli applied to the crus, presumably to the pyramid, which were characterized by a sudden relaxation and also inhibited the rubral tonicity. These phenomena all seem to point to the conclusion that we are dealing in the reticular formation with the mechanism involved at least in the associated movements seen in hemiplegic conditions, as pointed out by Walshe, as well as the tonic movements and postural conditions pointed out by Wilson. They do not, however, it seems to the writer, exclude the view that the above forebrain neurone systems may convey to the reticular formation impulses which result in purely kinetic reactions—rapid movements. The above stimuli were applied to a reticular formation apparatus already made hypertonic by decerebration and consequently not in a physiological condition suitable for observation of the normal effect of the above forebrain impulses acting upon it. Then, too, the "diagonal" limb flexions and extensions observed by Brown have much more significance as parts at least of a locomotive (kinetic) rather than a purely postural apparatus. In considering this question it is obvious that we are confronted with two aspects of the neural reactions: their *pattern* and their character as to movement and posture. It seems simpler to suppose that, in part at least, reactions having the same pattern involve common central efferent neurone systems, the tonic and kinetic differences being more due to the impulses acting upon such lower central motor mechanisms. From a histological point of view the character of the large "motor" cells of the reticular formation cannot be regarded as indicating a tonic function, for the same characteristics are possessed by the cells of Betz. They are probably best regarded, for the present, simply as long neurones conveying to distant lower motor neurones the results of neural influences integrated upon their bodies and dendrites.

The influences which bring out to some extent the latent motor pattern in the reticular formation when in the partially decerebrate condition of hemiplegia (the integration with the pyramidal system being, as above discussed, at a lower level) are well known. They are voluntary effort, emotion and, at times, attention. The mechanism of the "abnormal associated movements" thus brought out has been well discussed by Walshe (*Brain,* Vol. xlii, Part I). The path pursued is not entirely clear but it would seem to be probably the same path pursued when there is the same tendency, in less degree, to associated movement in normal conditions. Walshe apparently thinks it to be a proprioceptive reflex aroused by the voluntary movements of the normal side. Is it not also possible that the postural fixation represented by these movements are initiated by descending extra-pyramidal forebrain impulses and that the movements in question on the paralyzed side are so initiated? This might seem to be the case in synchronous bilateral associated movements, such as the "yawning" extension one (pandiculation), but the consecutive character of associated fist-clenching, in hemiplegia, would seem to favor Walshe's idea of a crossed proprioceptive reflex evoked by the movement on the sound side. We are confronted with the same dilemma in the case of the ipsilateral associated movements such as those seen in the case presented in this paper. A crossing via the corpus callosum would be one of the possibilities if the view is taken that a forebrain impulse accounts for the movement on the paralyzed side; but it must be remarked here that usually, in the case of associated fist-clenching, clenching the sound fist may inevitably produce a clenching on the paralyzed side, but attempted clenching of the paralyzed hand will not produce fist-clenching on the sound side. This is not conclusive, however, inasmuch as there may be, and probably is, an accession of tone also on the sound side but insufficient to produce results without the addition of pre-existing hypertonus.

The effect of emotion on the mechanism of posture and abnormal associated movements in pyramidal lesions also has its counterpart in normal activity. The effect of emotion on posture, expression and its tendency to produce a general increase of muscular tension is familiar. While the irregular defense reactions to certain stimuli, seen in lesions of the nervous system in thalamus and elsewhere, may be "release" phenomena due to defective cortical innervation, this hardly appears to be the case in the increased tension seen in intensive effort with emotion. The increase of associated movements, the rigidity of certain parts and the distorted facial expression seen, for example, in snapshots of runners breasting the tape in their final effort in a race, can hardly be called release phenomena. It is rather the calling out by the cortex of all available sources of energy including those tapped by the visceral system in ways made familiar to us in such work as that of Cannon and others. Here again we must apparently seek for forebrain paths other than the pyramidal one.

We finally come tq the curious phenomena exhibited in Wilson and Walshe's cases of "tonic innervation," in myotonia and, it seems to the writer, in the case here presented, i.e., cases where there is a difficulty, for example, in releasing voluntarily an object grasped in the hand. The effort simply causes a greater contraction of the grasping muscles although the releasing muscles are also contracted. The effort to release increases the difficulty and release is best brought about by diversion of attention. The difficulty is also increased by emotion. It may be added here that another case which was first thought to be functional but later it was decided to be a case allied to the dystonias shows the same effect of attention and effort but in a more isolated manner. The case is one of peculiar posture in a boy, the abnormality being markedly increased when attention is paid to it. The present case is allied, it would seem, to the athetosis group and also to the ordinary hemiplegic spasm but the peculiar semi-voluntary control makes it unusual. Here again the activities involved are obviously under at least partial cortical control.

From what has been previously said it would appear that the cortical paths available as a source for the above mentioned cortical influences are, besides possibly a partially intact pyramid, the pallio-pontile paths, cortico-thalamic paths and cortico-rubral and possibly other cortico-reticular paths, not to mention the obscure central tegmental-olivocerebellar path.

The general anatomical interpretation of the pallio-pontile paths is quite clear. It is obvious that in voluntary movement the proper cooperation of cerebellar activities must be secured; that they must for example, be either inhibited or increased either *in toto* or in part. It is surprising, however, how little that is concrete is known of these massive tracts. One clue is afforded by the abovementioned observations of Weed which seem to show that the frontal pallio-pontile tract, at least, inhibits certain tonus-maintaining cerebellar activities. This might furnish a partial explanation of the "tonic innervation" phenomenon—it would be due to a lesion preventing this inhibition of the tonic hemiplegic grasp, the "dead man's grip" as it was once picturesquely termed by a patient who, however, being a coachman, found he could make some use of it in holding the reins.

The above explanation, however, raises difficulties. Why, for example, do we find stimulation of the pyramid also producing such an inhibition of tone? We also encounter the phenonenon previously alluded to and already discussed by Bergmark, Head, Walshe and others, of a flaccid upper motor neurone paralysis, as well as the usual phenomenon of a spastic paralysis beginning as a flaccid one and the rare phenomenon of a spastic paralysis becoming flaccid. We also need data concerning the lateral (temporoparietal) pallio-pontile tract. It may be that these descending systems simply represent some form of influence proceeding from the cortical mechanisms of the three great groups of sense-organs especially concerned in maintaining posture—muscle-tendon (frontal), eye and ear, or it may be they are qualitatively different. It would seem not unlikely that they may have something to do with the cortical unconscious or semi-conscious maintenence of posture, especially of standing, but this is difficult to reconcile with the relaxing influence obtained by stimulation. In connection with this, however, account must be taken of the apparently double, excitatory and inhibitory, nature of the cerebellar activities according to some of the investigations named above, e.g., the phenomenon that apparently one portion of the cerebellum, that apparently receiving part of the frontal pallio-ponto-cerebellar path when stimulated diminishes tone while the cerebellum as a whole apparently maintains it. How far individual motion or tonus combinations are carried down in this path, as its size would appear to indicate, is another question of interest.

The pallio-rubral system perhaps does not rest upon so firm an anatomical basis as the pallio-pontile, but its function would, of course, reasoning anatomically, be the direct regulation of the rubral activities. It would seem that this system might have something to do with the influence of effort on posture and movement as seen in hemiplegia if a cortical original is postulated as suggested above. This suggestion, however, apparently conflicts with the evidence that seems to assign such a function to the globus pallidus. If it does act upon the lower mechanism of associated movement, its integrity would account for the ability of the patient indirectly to open her hand. Another view, not at all novel, is that it may in part perform those voluntary movements of axial musculature which do not appear to be so severely affected in pyramidal lesions. The position of the rubro-spinal tract in the cord does not, however, favor this view.

That the cortico-thalamic system should exert an influence on the emotional reactions, or rather constitute a part of its efferent mechanism, would appear probable and would explain some phenomena of pseudobulbar palsy, but here we are confronted with anatomical difficulties. We have no reliable descending thalamic path and must resort to some possible connection with subthalamus or hypothalamus or utilize the corpus striatum which probably receives thalamic fibers. The writer and others have already pointed out that the latter possibility may have a measure of truth, for many of the abnormal movements due to lesions of the caudate or putamen, according to Vogt's work, have a distorted resemblance to normal emotional reactions. There are again objections from comparative and other grounds, however. Apropos of the view as to the production of these abnormal involuntary movements by a lesion of the "inhibiting" putamen or nucleus caudatus it may be remarked that such a view apparently requires two different paths to the striatum, one the inhibiting path and another for the production of the movements in question. Such paths need not, however, be distinct outside the striatum, i.e., the afferent striatal path may have a shorter circuit in the pallidus and a longer one utilizing the short neurones of the putamen and possibly the caudatus, the latter being really coordinating and its removal causing the irregular, rather purposeless movements in question.

The question as to the relation of the splanchnic systems, both central and peripheral, to tonus, and especially to the peculiar rigidity seen in striate lesions, cannot be discussed here but it is obviously the thalamo-pallial affective reactions which have the closest relation to the splanchnic system.

As Walshe observes, the question of the production of the abnormal involuntary movements is the most difficult of all. His comment, anent the attribution by Holmes of tremors and rigidity seen in midbrain lesions to lesion of the cerebello-rubro-spinal path, that "if this path be the efferent link, as we have supposed, of the proprioceptive reflex arc, it is surprising that its interruption should be followed not by atonia and ataxy, but by tremor and rigidity," expresses a thought that has also occurred to the writer. Certain points may be noted, however: first, that only one of the cerebellar efferent paths is affected; second, that in cerebellar lesions tremors, including an intention-like tremor, have been described; third, that the lesions in question usually involve the nucleus ruber, as well as the adjoining reticular formation, and where this nodal point is involved the picture may very well change. It must be remembered, however, that "choreo-athetoid" movements have been described in superior peduncular lesions. Descriptions, however, of abnormal movements are greatly lacking in uniformity and precision.

It may be remarked here that the tremors and abnormal involuntary movements in general are also markedly influenced by other activities of the nervous system and that some of these activities such as voluntary movement and effort, as well as emotion, are of cortical origin. Consequently much of the above discussion as to associated movements applies here also.

That various dissociations of influences normally integrated will account ultimately for many or all abnormal involuntary movements seems probable, but when the number of integrative levels is considered it is seen that the problem is complex. It would seem probable that in the case of abnormal involuntary movements the "disintegrations" are probably on the efferent side while the true ataxias are more in the afferent proprioceptive side including coordinating proprioceptive mechanisms such as the cerebellum.

The above discussion has not been made with the hope of clearing up a subject both difficult and requiring more concrete investigation and observation, but is simply a slight attempt to correlate some of the phenomena rather more definitely with anatomical structures and to show how really limited our knowledge, anatomical, physiological and clinical, is regarding many important relevant points and how impossible it is to draw conclusions without being confronted with alternatives between which we cannot choose in the present state of our knowledge.

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THE STATIC OR POSTURE SYSTEM AND ITS RELATION TO POSTURAL HYPERTONIC STATES OF THE SKELETAL MUSCLES*

SPASTICITY, RIGIDITY AND TONIC SPASM

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In a previous paper,[1] I have presented at some length the evidence in favor of a dual efferent system. According to this view, motility is subserved by two components, each represented throughout the entire efferent nervous system by separate neural mechanisms, which are physiologically and anatomically distinct. This is true not only of the segmental or reflex nervous system but also of the brain and cerebellum.

Under normal conditions these two systems act in unison, and their harmony of action is only disturbed by the accidents and dissociations of disease.

One of the components is the movement proper, which is subserved by what I term the *kinetic system* (motion system).

The other represents that more passive form of contractility which we recognize in tonus, posture, attitude and equilibrium. This posturing or static function of the efferent system is, I believe, subserved by separate neuromuscular pathways, which I term the *static system* (posture-system).

The term static is used here to designate that peculiar property of the striated muscle fiber by which it becomes fixed in posture. In the reflex nervous system this is manifested by postural tonus and at the higher levels by the various postures and attitudes. In muscle physiology this function is represented by the so-called "lengthening and shortening reactions." This intrinsic property of muscle tissue is dependent on postural tonus, by which a muscle may be lengthened or shortened during contraction without corresponding changes of tension. This alteration in postural length without corresponding changes of tension facilitates the easy transition from posture to movement which characterizes motility.

There are many reasons for the assumption that the kinetic system is related to the anisotropic discs and the static system to the sarcoplasm of the muscle fiber as these elements differ in structure, innervation, mode of contractility and metabolism.

There are also reasons for the assumption that the transformation of movement into posture is effected by fixation of the sarcoplasm which thus converts the contractile muscle fiber from a kinetic into a static mechanism. It is likely, however, that each system participates to some extent in the other function. And while the essential function of the static system is postural it is also contractile and therefore to some extent kinetic—and the kinetic system while essentially motional also plays a certain rôle in static or postural function.

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*Amplification of a discussion on Decerebrate Rigidity at a clinical conference of the Neurological Department of Columbia University.

According to this conception, then, movement is subserved by a kinetic, and posture by a static mechanism, the two systems supplementing one another and working together in harmony. For every movement starts from posture and terminates in posture, posture following movement like a shadow. At the same time, the postural mechanism would exercise a stabilizing and steadying influence upon the course of movement itself.

The kinetic system is concerned with the active movement; the static system is concerned with what might be termed the passive form of movement as expressed in postures and equilibrium. Both are manifestations of the efferent mechanism but differ in the character of their function, although subserving a common end—motility.

In the present discussion, I shall limit my remarks to certain aspects of this postural function, which includes such disorders of muscle tone as the spastic state, the muscular rigidity of paralysis agitans, decerebrate rigidity, and the tonic or postural spasms.

Motility as it is observed in animal life may be divided more or less arbitrarily into three distinct groups, viz.; reflex, automatic-associated, and isolated-synergic or dissociated types of movements.

All of these forms of movement are intimately associated with that other component of motility which we term posture. And the same postural groups may be recognized as in the classification of movement, viz.; *reflex postures, automatic-associated postures* and *isolated-synergic types of posture* corresponding to their respective motion mechanisms. And it may be stated as a general principle that there is no form of motility from the simple reflex to the most skilled and individualistic type of movement which functions without a corresponding static or posture mechanism.

ANATOMICAL CONSIDERATIONS (THE STATIC SYSTEM)

In a general way, the central nervous system may be said to represent three great structural and functional divisions. There is the segmental nervous system which contains the great reflex systems of the neuraxis, and is the archaic representation in man of nervous reactions of the lowest forms of life. The other is the paleo-encephalon represented by precursors of the corpus striatum and optic thalamus which constitute the higher coordinating sensory and motor mechanisms of the lower forms of life. The neo-encephalon represents the latest stage in the evolution of the nervous system and has its greatest development in man. It subserves the highest functions of motility and sensibility.

These three great functional divisions of the nervous system are related to the three great types of movement and posture already mentioned.

In this rather schematic division of motility and its mechanisms it should be understood that the development from one type of movement to another and from one structural division to another is a very gradual one—a transition of structure and function which may only be traced in comparative anatomy and physiology but the imprint of which remains stamped on the central nervous system of man.

The segmental nervous system is the seat of the various reflex movements and reflex postures. The other types of movement and posture are localized as follows.

In previous studies of paralysis agitans,[2] I expressed the view that the corpus striatum is a paleokinetic center for the control of "automatic and associated movements," and the Rolandic area of the cerebral cortex is a neokinetic center controlling the isolated-synergic movements of cortical origin.

Their various postural functions are, I believe, subserved by systems which are a part of the cerebellar mechanism.

According to this view, paralysis agitans is produced by disease of the efferent *pallidal system* of the corpus striatum and is characterized by paralysis of automatic-associated movement. Spastic paralysis is produced by disease of the *pyramidal system* of the cerebral cortex and is characterized by paralysis of isolated-synergic movement. Each form of palsy—*pallidal* and *pyramidal*—is accompanied by a special form of muscle hypertonus with characteristic attitude and posture.

The essential integrating and correlating mechanism for the control of static motility I believe to be the cerebellum. Afferent impulses from the periphery and efferent impulses from the cerebral cortex pass to this organ before their final distribution by way of the cerebello-spinal system to the posturing or static mechanism of skeletal muscles.

The cerebellum presents phylogenetic evidences of two great divisions, a paleocerebellum and a neocerebellum, which correspond to similar divisions of the cerebrum. The paleocerebellum receives impulses directly from the spinal cord, which pass to its central portion and constitute the vermian system. The neocerebellum receives impulses from the cerebral cortex, which pass by way of the pons varolii to the cerebellar hemispheres, and constitute the hemispheric system.

The older cerebellum stands in relation to what may be termed the *paleostatic system* which controls the older static or postural functions of "automatic and associated type." It takes its origin in the older nuclei of the vermis cerebelli (N. Fastigii, globosus, emboliformis). The automatic-associated posture of standing, as observed in decerebrate rigidity, would be one of the functions of this paleostatic system.

The cerebellar hemispheres, on the other hand, regulate the higher postural functions of motility through the medium of a neostatic system, which takes its origin in the cells of the dentate nucleus. The neostatic system is controlled from the cerebral cortex by special tracts which connect the various regions of the cerebral cortex (frontal, parietal, temporal and occipital) with the dorsal and ventral nuclei of the pons varolii, and thence to the opposite hemisphere of the cerebellum.

In posture, as in movement, no sharp line of demarcation can be drawn between the various types, and one group merges imperceptibly into another. It is probable, I believe, on phylogenetic grounds, that the parieto-temporo-occipital tract to the dorsal pontine nuclei is older than the fronto-pontine system which descends from the frontal lobe to the ventral nuclei of the pons. The latter system I regard as the homologue in the static or posture sphere of the cortico-spinal or neokinetic system. The fronto-ponto-cerebellar or *neostatic system* descends in the anterior limb of the internal capsule and mesial portion of the cerebral peduncle in close relationship with the pyramidal tracts, and it is interesting to note that both structures, the one subserving the neostatic and the other the neokinetic functions of motility, receive their myelin sheaths subsequent to birth.

The other cortico-cerebellar systems (parieto-occipito-temporal systems) pass in the lateral portion of the crus cerebri to the pontile nuclei, and thence to the opposite cerebellar hemisphere. It is my belief that they represent cortico-cerebellar connections subserving higher automatic associated types of posture in connection with the various sensory areas of the cerebral cortex and are the homologues in the static sphere of cortico-thalamic connections with the strio-spinal or paleokinetic system.

The existence of these intermediate kinetic and static systems connecting the various sensory areas of the cortex with the thalamo-striate mechanism on the one hand (cortico-paleokinetic systems), and the cerebellum on the other (cortico-paleostatic systems), are confirmatory of my view of the parallelism of structure and function which underly the development of motion and posture.

The chief efferent systems of the cerebellum (*paleostatic* and *neostatic*) pass by way of the superior peduncles to special cellular divisions of the nucleus ruber. The paleocerebellar system terminates in the nucleus magnocellulatus and the neocerebellar system in the nucleus parvicellulatus. From here they descend in the rubro-spinal and other systems of the spinal cord, then passing to the sarcoplasm of the skeletal muscles as peripheral static systems. The distribution of the static system in pheripheral nerves and its mode of termination in the muscle fiber is still one of the mooted questions of histology and physiology. There is already considerable evidence in favor of the dual innervation of the striated muscle fiber but anatomists differ widely as to the manner in which these peripheral static or posture systems reach the muscles.

When, however, one considers the well developed central mechanisms for the dual function of the efferent system, there can be but small doubt as to the existence of a corresponding difference of structure and function in peripheral nerves. Ransom has already demonstrated, by a special method of staining, the existence of large numbers of non-medullated nerve fibers in peripheral nerves, and perhaps here is concealed the solution of this intricate question.

It is interesting to observe that all the posture systems, neostatic as well as paleostatic, pass to the cerebellum for final integration and coordination. This is in accord with the nature of the posturing mechanism and its secondary and unconscious rôle in motility. For while the higher forms of movement are initiated as conscious and voluntary processes, the corresponding postures are secondary, and follow automatically in the path of movement. This explains, to my mind, why the neostatic system does not pass directly to the gray columns of the spinal cord, as do the pyramidal tracts (neokinetic system) but is directed first to the cerebellum, which is the great central correlating mechanism for posture function.

Motion is an active conscious process, while posture is a passive unconscious process which becomes automatically operative as soon as movement ceases. It is for this reason that the integration and coordination of posture takes place in an organ like the cerebellum, at the unconscious level of activity, in contrast to neokinesis which is subserved by a direct cortico-spinal connection.

This in brief is my conception of the static system and its relation to motility. Let us now consider some of its relations to symptomatology.

THE STATIC SYSTEM AND POSTURAL HYPERTONICITY OF THE SKELETAL MUSCLES

As we have seen, there are as many types of posture as there are of movement, and with corresponding development of neural mechanisms. We must therefore recognize the existence of special fibers in the cortical, capsular, crural, cerebellar, pontile, bulbar, spinal and peripheral divisions of the nervous system to carry out this static or postural function, corresponding to homologous systems for the kinetic or motorial function.

When some portion of the kinetic system is injured, paralysis immediately results; if the injury takes place in the central nervous system the lower motor centers are released from control, and a characteristic postural rigidity develops. This peculiar over-action of certain muscles takes place in response to the principle that destruction of a higher center releases the lower kinetic mechanisms from control. When this has taken place, as after all other forms of movement, the motion-complex becomes fixed in posture and we have produced that curious paradox of a paralyzed center above and a functioning center below, which has expressed itself in terms of *postural-hypertonus.*

At the present time, two great motion sys-
terms of *postural hypertonus.*
neokinetic systems. Destructive lesions in the course of either system would release subordinate motor centers from control which then discharge in terms of motion-formulae and become fixed in terms of posture-formulae.

These are the factors with which we are more particularly concerned in any study of postparalytic hypertonicities of the skeletal muscles.

TYPES OF POST-PARALYTIC POSTURAL HYPERTONICITY

Many different types of postural hypertonus following paralysis may be recognized, which depend upon the nature and localization of the lesion within the central nervous system. The more important of these are the spasticity of pyramidal tract disease; the muscular rigidity of paralysis agitans; the decerebrate rigidity of midbrain section, and the postural hypertonic effects of transverse lesions of the pons, medulla and spinal cord.

Cortical or Pyramidal Types of Postural Hypertonus (spasticity).—One of the most characteristic symptoms of organic disease of the central nervous system is spasticity, or spastic hypertonicity of the skeletal muscles.

The affected muscles while weak or paralyzed are in a state of overaction and postural fixation which is dependent upon the overactivity of subordinate kinetic and static centers.

Any injury of the neokinetic or cortico-spinal system is followed by paralysis of the isolated-synergic movements of cortical origin and release of the lower motor mechanisms of the spinal cord. This is commonly referred to as a loss of cerebral inhibition. In spasticity the kinetic and the static systems are both in action. There is a kinetic discharge which is accompanied by postural fixation. The characteristic attitude and postural deformities of the purely spastic type of hemiplegia are therefore caused by postural hypertonic fixation resulting from paralysis of the cortico-spinal pathways (pyramidal system), and the overactivity of the corresponding spinal mechanism.

Paralysis of the cortical center for neokinesis releases the subordinate neokinetic spinal mechanism which is especially concerned with this type of motion.

This is the reason, I believe, that the chief postural deformities of the spastic state are so sharply localized in the distal portion of the extremities, and more especially the hands and fingers. For the hand is the chief instrument and special localization of the higher neokinetic activities of man, and therefore shows the greatest degree of palsy in hemiplegia of the pyramidal type.

The tendency to clonus, the exaggerated tendon and periosteal reflexes in spastic states, I would also explain by the heightened reflex activities of the spinal cord. The cortico-spinal system passes directly to the reflex mechanism of the spinal cord in the interest of the dissociation of movement, and any diminution or loss of this inhibitory control releases the neospinal mechanism, which explains the heightened reflex activity of tendon and muscle response.

A further elaboration of this question will be found in my earlier study of neokinetic and paleokinetic representations in peripheral nerves.[8]

Striatal or Pallidal Types of Posture Hypertonus (paralysis agitans rigidity).—The rigidity of paralysis agitans is a postural hypertonicity resulting from paralysis of the pallidal system of the striospinal pathway. In my previous studies of paralysis agitans this point of view has been presented, together with evidences showing that the efferent motor system of the corpus striatum (pallidal system) represents an internuncial common pathway for the passage of motor impulses for automatic-associated types of movement (paleokinetic system).

The characteristic attitude and postural deformities of this affection present many points of difference from those associated with the spastic state. There is the mask-like expression of the face, the peculiar attitude of head, trunk and arms, and the characteristic position of the hand and fingers. The distribution of the palsy and associated muscular rigidity differ essentially from that observed in the spastic state.

Furthermore, the hypertonicity is more of the rigid waxy type, and lacks those special elements which characterize the spastic state. This I would explain by the peculiar location of the lesion in the pallidal system of the corpus striatum. Paralysis of this system releases from inhibition the various kinetic centers of the midbrain and hypothalamic region, among which may be mentioned such structures as the red nucleus, corpus subthalamicum and the substantia nigra. When these subordinate centers of the paleokinetic system are released, their various motion-complexes are discharged and become fixed in postures (postural hypertonicity).

It will be recalled, however, that the hypertonicity of paralysis agitans is not accompanied by clonus and other evidences of exaggeration of the reflex activities. The reason for this is obvious. The lesion of paralysis agitans releases only the *kinetic centers of the midbrain and hypothalamic region,* and the lower reflex centers of the spinal cord are still controlled at this higher level. Whereas in the case of the corticospinal pathway (pyramidal tracts) all inhibition is removed from the lower spinal mechanism, with the development of the characteristic reflex activities of the spastic state.

Another point of interest is the tremor of paralysis agitans, which is so characteristic of the pallidal type of palsy. This tremor is also the expression of subordinate kinetic centers of the hypothalamic and midbrain region which are under the control of the corpus striatum, and which when released express themselves in terms of tremor. It is not unlikely that the striatal connections with the red nucleus are closely related to this form of tremor; but here we are still in a very conjectural field. It would appear probable, however, that the red nucleus in addition to its static component, which stands in relation to the cerebellum, has also a kinetic component which is under the control of the corpus striatum.

Pallido-Pyramidal Types of Postural Hypertonus (spastic rigidity).—The proximity of the corpus striatum (pallidal system) to the internal capsule (pyramidal system) is such that simultaneous involvement of both structures is not uncommon. Indeed, in vascular lesions of the basal ganglia, it is probable that pure forms of either pyramidal or pallidal palsy are quite rare and that a combination of the two types is the more common.

Involvement of both systems, the pallidal and pyramidal, produces a *spastic-rigid* state which combines in varying degrees the characteristics of both types of postural rigidity.

Combined or Pallido-Pyramidal Type of Hemiplegia.[4]—In the combined or pallido-pyramidal type of palsy there is not only loss of isolated-synergic movements of cortical origin, but also of the automatic-associated movement, referable to the striatal mechanism. With this combined form of palsy there is also a corresponding fusion of their postural elements which to my mind explains the variety of postural types in the ordinary vascular hemiplegia.

The special feature of this fusion of hypertonicity is the combination of the waxy rigidity of paralysis agitans and the characteristic symptoms of the spastic state.

Indeed, all degrees of variations may be observed in the combined types of hemiplegia, depending upon the relative loss of the two great types of movement, automatic-associated and isolated-synergic, and the corresponding postural changes.

Midbrain or Paleocerebellar Type of Postural Hypertonus (decerebrate rigidity).—Another type of hypertonicity which has awakened considerable interest and investigation, is the decerebrate rigidity of Sherrington. This is a peculiar state of muscular rigidity which corresponds to the automatic-associated posture of standing and is produced by a section through the midbrain at the level of the anterior colliculi.

A mesencephalic transection at this level severs the pyramidal tracts (neokinetic system), the pallidal tracts (paleokinetic system), as well as the cortico-ponto-cerebellar tracts (neostatic system). The various subordinate centers of the pons, medulla and cord, both kinetic and static, are released and the skeletal muscles assume the postural rigidity of standing which is characteristic of this level.

As the older static system of the cerebellum (paleocerebello-rubro-spinal system) for the control of automatic-associated types of posture is still functionating, retaining its afferent connections with the spinal cord and the efferent with the red nucleus, the automatic posture of standing, characteristic of decerebrate rigidity, is maintained. When the section is carried below the red nucleus, which severs the paleostatic system, the typical decerebrate posture no longer appears and the other types of postural reaction make their appearance (pontine and bulbar). Decerebrate posture is also abolished by section of the superior peduncles of the cerebellum, which would sever the connection between the efferent cerebellar system and the red nucleus.

Decerebrate rigidity may, therefore, be regarded as the postural expression of paleostatic activity, which develops after the elimination of cortical and striatal inhibition.

Spinal Types of Postural Hypertonus (paraplegia in flexion).—A transverse lesion of the cord would sever all of the long projection systems, both static and kinetic, and release its various intrinsic spinal reflex mechanisms. In response to this loss of the higher inhibitory control, the characteristic reflex of flexion develops which becomes fixed in postural hypertonus, the extreme expression of which is "paraplegia in flexion."

An extensor type of spinal paralysis is also encountered (paraplegia in extension). This form is characteristic of pyramidal tract disease and is merely the spinal expression of the *pyramidal* type of hypertonus, as observed in purely spastic states.

COMMENTS

When we consider, therefore, that every type of central palsy, both neokinetic and paleokinetic, is followed by postural hypertonicity, and when we also consider that there is evidence of a static system for the control of posture throughout the entire central and peripheral nervous sytems, the application of experimental observations on postoperative hypertonus in animals to clinical observations in man should be used with considerable caution.

This is especially true of experimental transections of the brainstem (crus, pons and medulla), as these structures include the various *prespinal centers of motion and posture* (motion centers and posture centers), and is one of the most complicated in the realm of neuro-physiology and neuro-anatomy.

For here are congregated, according to my conception, groups of centers subserving *motion formulae* under the control of the corpus striatum, as well as groups of centers subserving *posture formulae* under the control of the cerebellum.

Kinnier Wilson[5] in a recent and very suggestive study of decerebrate rigidity in man introduces the subject as follows:

"Briefly, the type of case herein described and analyzed is one in which there is evidence of withdrawal of cortical control in the form of unconsciousness or semi-consciousness, the result commonly of cerebral hemorrhage—submeningeal or intraventricular or both—or of meningeal inflammation, or from the effect of certain intracranial tumors; also, for that matter, of hysteria. Coupled with this impairment of consciousness has been the appearance of tonic rigidity, to which frequent exacerbations in the form of tonic or postural fits, accentuating and exaggerating the background of tonic posture, are superadded.

"It will be shown, further, that in certain types of organic and functional disease these postural attitudes may be adopted involuntarily with full consciousness, and that partial or fragmentary types also occur. To this last statement much importance is attached, since the hypothesis to be subsequently developed is that the involuntary postures of a limb, or segment of a limb, in many forms of nervous disease, as well as the transient positions assumed in involuntary movements such as athetosis, are in reality simply parts of the complete decerebrate attitude."

In the first place one might ask, Is it definitely known whether mesencephalic transection would produce exactly the same symptoms in man as in the lower mammals? The effects of ablation of the cerebral cortex, section of the pyramidal decussation and spinal cord transections differ in their effects on animals and man. And while there is no doubt that the tonic spasms and postural rigidities of the *decerebrate type* occur as a result of lesions in the neighborhood of the cerebellum and red nucleus in man, to ascribe nearly all manifestations of postural hypertonus to this mechanism represents an attempt to cast a very extensive group of symptoms in much too narrow a mould.

Furthermore it is only rarely that lesions are observed so strictly localized as to reproduce mesencephalic transection in man. A very significant fact in Wilson's group of cases presenting supposed decerebrate symptoms is the rather diffuse character of the lesions in many of them. There are also included such clinical conditions as cerebral diplegia, infantile hemiplegia, microcephaly with diplegia and athetoid attitude, as well as cases of chorea and athetosis, all as presenting partial or fragmentary signs of decerebrate rigidity, in the sense in which the term is used by Sherrington.

That postural symptoms, both general and local, more or less suggestive of experimental decerebrate rigidity are also encountered in man is unquestionable, but that these are all to be referred exclusively to the midbrain level is, in my opinion, more than doubtful.

In the first place I would recall that the decerebrate posture or attitude is of the automatic-associated type and represents a fixation in terms of posture of a well-known automatic-associated act.

As the corpus striatum is the essential organ for the correlation of automatic-associated types of movement, a release of this mechanism, as occurs in such disorders as chorea and athetosis, would liberate movements of this type and among others those suggestive of the decerebrate postures of the extremities and trunk. Furthermore, in the cortical or *pyramidal* types of palsy with loss of isolated-synergic movements, there are present numerous *associated movements,* many of which emanate from the striatal mechanism. Certain of these movements of the automatic-associated type when localized in the hand and arm may resemble or suggest a true decerebrate posture of the extremity. Properly speaking, these are kinetic and originate in the strio-spinal mechanism, and are quite different in their symptoms from the somewhat similar manifestations of decerebrate rigidity.

Therefore in pyramidal tract disease, with preservation of automatic and associated movements (diplegia), and in such disorders as chorea and athetosis, the appearance of a *decerebrate* symptomatology may be regarded as spurious and referable to the striatal mechanism.

This, however, does not exclude the existence of the true decerebrate rigidity in man in association with lesions in the region of the midbrain and cerebellum, and referable to the *paleostatic system.* This decerebrate manifestation in its pure form, as Wilson himself admits, is quite rare and great caution should be used in its more general application.

The views which have been expressed may be briefly restated as follows.

The function of motility is subserved by two distinct components, which express themselves in movement and in posture. The active component is movement, which is served by a kinetic or motion system.

The more passive component is posture, which is served by a static or posture system. Both systems, I believe, are represented throughout, from the lowest to the highest level of the efferent neuro-muscular mechanism.

The reflex nervous system is concerned with reflex movement and reflex posture, the paleokinetic and paleostatic nervous system with automatic-associated types of movement and posture, and the neokinetic and neostatic with isolated synergic types of movement and posture.

The essential correlating mechanism of all static or postural activity is centered in the cerebellum, which is under the control of spino-cerebellar and cortico-cerebellar pathways. There are two forms of cerebellar activity, paleocerebellar and neocerebellar, which correspond to different types of posture.

The neostatic function of the cerebellum is under the control of a cortico-ponto-cerebellar system (fronto-pontine tracts).

The paleostatic function of the cerebellum is under the control of the spino-cerebellar system. The efferent cerebellar systems pass to paleostatic and neostatic portions of the nucleus ruber, as well as other prespinal posture centers, from which they descend the spinal cord to the peripheral static systems.

According to this conception the static system, like the kinetic, is represented in the central nervous system by neostatic and paleostatic systems which subserve the older and more recently acquired forms of posture. This is in harmony with the theory that I have expressed, that motion and posture develop side by side as twin mechanisms. When there is movement there is also posture, and when a kinetic mechanism is formed, a static mechanism is also formed, a parallelism which may be traced in the whole field of motility both functional and structural.

Two fundamental types of central palsy may be recognized. A neokinetic or *pyramidal type* characterized by paralysis of isolated synergic movement, and a paleokinetic or *pallidal type,* characterized by paralysis of automatic-associated movement. Both are associated with characteristic postparalytic types of postural hypertonus.

The *pyramidal type* of *postural hypertonus* is the well known spastic state. The *pallidal type* of *postural hypertonus* is the characteristic postural rigidity of paralysis agitans. Combinations of these types are frequent, from involvement of both the corpus striatum and internal capsule, giving rise to *pallido-pyramidal types* of *postural hypertonus.*

A rare form of postural hypertonus in man is the midbrain or paleostatic type, which is characterized by the postural hypertonus of decerebrate rigidity. In its purest form it is produced by lesions in the region of the midbrain and cerebellum.

This type, according to Kinnear Wilson, is characterized by retraction of the head, opisthotonos, extension-pronation of the arm, flexion of the wrist, and extension of the legs and feet. It is a replica in man of the posture of experimental decerebrate rigidity in animals.

In this postural manifestation there is loss of the pyramidal and pallidal functions of motility with preservation of the paleokinetic and paleostatic functions of the midbrain and cerebellum, of which the automatic-associated posture of standing is a part.

The spinal types of postural hypertonus are dependent upon release of all the reflex spinal mechanisms and in man are characterized by the flexion type of spinal paraplegia.

All of these various postparalytic types of postural hypertonus are indications of overactivity in subordinate motion and posture centers following paralysis and loss of the higher mechanism.

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AN EXTENSIVE SPINAL ARACHNOID FIBROBLASTOMA

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This case is presented because it illustrates a rather unusual type of spinal cord tumor which was incompletely removed on two occasions. When the patient first presented himself at the hospital one year after the onset, he was unable to walk; but after operation normal function returned to the limbs. The paralysis gradually developed again after eight years when he was again relieved by a second operation. Three years later there was a third recurrence necessitating a third operation, which, however, the patient did not survive.

Patient.—Mr. C. T. Age twenty-nine at the time of his first admission to the Presbyterian Hospital in September, 1909. He was married. His occupation was that of an architect.

History.—In October, 1908, while playing a game of football, the patient was hurt in the scrimmage, immediately after which he felt an intense pain in the region of the spine between the shoulder blades. The pain was so severe that it was impossible for him to remove his clothes without assistance. It continued for a week and disturbed his sleep, but after this time gradually subsided. Ten days later he entered a broad jump contest, but upon attempting to jump, suddenly lost power in both legs and fell. His legs did not become stiff, but he could not move them as rapidly as before. He noticed a numbness in his toes apparently without any actual loss of sensation in this region. Gradually his legs became spastic and the numbness increased until September, 1909, when he presented himself at the hospital.

Previous History.—In childhood he had measles, typhoid fever and pneumonia. Otherwise his general health had been excellent.

Family History.—His family history as to nervous or mental disease was negative.

VOLUNTARY MOTOR SYSTEM. *Attitudes.*—Both legs were spastic and weak. Inspection found no sores or other abnormal marks upon his body.

Gait.—He walked with difficulty and moved his legs slowly.

Reflexes.—The knee jerks were very active. There were bilateral ankle clonus and bilateral Babinski. The cremasteric and abdominal reflexes were difficult to elicit.

Myasthenic Status.—There was increased tone of both legs but no atrophy. He had good control of both arms and the neck muscles. There was no complete paralysis in any part of the body.

GENERAL SENSORY. *Touch.*—The patient felt the touch of a finger anywhere on legs, back and abdomen.

Pain.—There was a numbness of both legs, extending up the trunk to a level an inch above the nipple. There were no shooting pains nor gastric crises. There was pain between the shoulders in the region of the original injury, and where he first felt pain after he was hurt. This pain was elicited by jarring or bending forward and had persisted ever since the injury. In some places on the legs and on the abdomen below the nipple it was difficult for him to recognize the prick of a pin other than as a touch. In other places he experienced pain when pricked with a pin.

Temperature.— He did not recognize heat or cold, presumably in the area below the line of sensory limit, sometimes calling the hot cold and the cold hot.

Supplementary Examination.—The patient was observed from time to time. The deep reflexes in the legs became more active; the sensory disturbances increased until pain and temperature sensibility were more and more impaired.

As to rectal and vesical control, his bowels moved daily without catharsis and he had no trouble with his sphincters. For the past five months he had had trouble with starting micturition. Just above the level of the nipple, a little higher on the left side, there was a well defined band three to four inches wide extending around the body, which was distinctly hyperesthetic. From this level down the sensation was disturbed as described above. The thoracic and abdominal organs and the cranial nerves were all normal and the arms showed no sensory disturbances or paralysis.

Clinical Notes.—The weakness of the legs increased daily. On September 10th and 13th lumbar punctures were made but no fluid obtained. On September 23d a third lumbar puncture yielded about 20 c.c. of light colored amber fluid, which came out slowly drop by drop, under low pressure. This fluid showed a marked positive Noguchi reaction and contained very few cells, mostly lymphocytes. During this time his temperature, pulse and respiration were normal. Urine was normal. Wassermann reaction of the blood was negative. There was no tuberculin reaction. The blood count showed red blood cells 5,120,000; hemoglobin 87 per cent; white blood cells 8,000; polymorphonuclears 63 per cent, lymphocytes 33 per cent, eosinophils 2 per cent, basophils none.

It was inferred that the patient was injured at the time of the scrimmage in football and that owing to that injury there was a subdural hemorrhage in the upper dorsal region of the cord and, perhaps, some laceration of the meninges, possibly a fracture of one of the vertebrae, although a radiograph of the spine showed no abnormality. It seemed likely that at the time of the injury there was not sufficient pressure on the spinal cord to cause any decided cord symptoms but that a lesion gradually developed which in some way compressed the cord and slowly increased the symptoms. During the month of October the weakness increased so that the patient was unable to walk.

Operation was advised and on October 27, 1909, Dr. Joseph A. Blake performed a laminectomy as follows: An incision from the seventh to the fourth dorsal vertebra was made. Considerable bleeding was met with superficially. No evidence of a fracture or dislocation in the vertebrae could be found. The dura, when exposed, showed a little old fibrinous exudate. On being opened the dura was at once expanded bilaterally by a tumor mass which resembled a large blood clot and which surrounded the cord from the seventh cervical to the third dorsal vertebra about 7.4 cm., and showed the consistency of a granuloma. The cord at this point was very much compressed so that it seemed smaller in diameter than above or below, but its continuity still existed. The tumor mass was removed piecemeal by a spoon and forceps, care being taken to preserve the cord. After as much as possible of this material was removed, the dura was sutured over the cord with fine sutures. The operation occupied one hour and five minutes. The patient made a fair recovery from the anesthetic.

Almost immediately after recovering consciousness the patient could move his legs much better than before operation. The temperature rose on the second day after operation to 102 degrees, but immediately fell to normal after the removal of a large bloody exudate which had collected beneath the skin at the point of operation. Catheterization was necessary on the night of the operation but not afterwards.

The patient's condition steadily improved. He could draw up his legs almost to full flexion, which had been quite impossible before operation. The pain and temperature senses improved so much that they were nearly normal. The ankle clonus, patellar clonus and Babinski reaction still continued.

November 28, 1900: The improvement continued. The wound was entirely healed. The ankle clonus and patellar clonus were diminished. The patient walked about with a chair and was gaining complete control of his legs.

December 22, 1909: The patient left the hospital walking very well without assistance and without guiding himself with a chair or cane. The sensory disturbance had disappeared. A slight ankle clonus still existed in the right leg and a slight Babinski reaction was present on both sides. He experienced no more pain between the shoulders.

Examination of the Tumor Tissue.—The tissue in the gross appeared like a blood clot. Microscopically it consisted of endothelial cells arranged in layers around the blood-vessels and lying in a meshwork of fine fibrils. In some places the cells presented a regular starlike radiation outward from a blood-vessel. This picture is very much like that of a neuro-epithelioma, in which the glia cells and fiber striations around the blood-vessels form stars. In other places the arrangement of the cells appeared more like that in a hemangio-endothelioma of the dura. Throughout the tumor tissue fresh hemorrhages and the remains of old hemorrhages were seen. The tumor was diagnosed as an endothelioma of the perithelial type.

Interval History.—After leaving the hospital the patient returned to his home. He was able to get about with a cane at first, and after steady improvement he could walk without one. After nine months he walked without limping and was able to play tennis. Running, however, was somewhat difficult, owing to weakness of the right foot. Pain in the lower lumbar region persisted three months after leaving the hospital. After two years he felt completely recovered and entered into all his former activities, including athletics. The only symptoms noticed at that time were tiring of the right leg after considerable exercise and occasional slight pain in the lower lumbar region.

Two years after this he was practically well, and remained so until in the fifth year after leaving the hospital (1913) he began to notice an increase in the frequency and severity of the lumbar pain, which came mostly at night. This was relieved by lying on his back with a pillow under his lumbar region. Occasionally the pain would come when the patient dived into the water or jumped on the ground landing hard on his heels, and also when bending over a draughting board. One year later the pain increased and he began to notice an increasing weakness of the right leg and numbness gradually spreading from the foot upward. There were no tingling, formication, heat or cold sensations, but a "heavy dead feeling."

Two years later (1915) the left leg became similarly involved and both legs gradually became worse, accompanied with pain in the back which frequently kept him awake at night. The pain was aching in character, never radiating. There was also occasional pain in the right side of the abdomen. No bands of increased or decreased sensation on the body were noted. Walking became more difficult due to increasing stiffness of the legs and clonus when he put the ball of the foot to the ground. The bowels were normal. Some difficulty in urination was noted on starting the stream. Sexual functions were normal.

May 17, 1917: The patient reentered the Presbyterian Hospital where the following examination was made:

General Examination.—There was a scar from the first thoracic spine to the sixth. The right leg appeared slightly smaller than the left and also slightly shorter. There was a slight drop foot on the right side.

Circumference of leg 15 cm. below patellar region, right 30.5 cm., left 32.5 cm.

Circumference of thigh 25 cm. above patellar region, right 48 cm., left 49 cm.

Palpation and massage showed nothing abnormal.

VOLUNTARY MOTOR SYSTEM. *Attitudes.*—There was difficulty in standing.

Gait.—When walking there was a general tremor of the body which seemed to be caused by clonus in both ankles. The patient walked in a slow, shuffling manner with a spastic gait. The difficulty in standing and walking was not increased when the eyes were closed. Special tests, such as walking on toes, on heels, backwards and sidewise, also the tests of Fournier, were performed imperfectly or with great difficulty because of the spacticity and weakness in the legs.

Coordination.—Equilibratory and nonequilibratory coordination were normal except in so far as the tests were interrupted by the spasticity and weakness in the legs.

Skilled acts were normal. There was no dyspraxia. Handwriting was normal.

Reflexes:

DEEP	*Right*	*Left*
Jaw jerk	1	1
Pectoral	1	1
Biceps	2	1
Triceps	2	1
Ulnar	2	1
Patellar	3	3
Suprapatellar	3	2
Achilles	4	4
Wrist	2	1
Radial	2	1
Periosteal	2	2
Periosteal crossed	1	1
Ankle clonus	4	4
Patellar clonus	0	0
Wrist clonus	0	0
Jaw clonus	0	0

SUPERFICIAL		
Supra-umbilical	2	1
Suprapubic	2	1
Upper lateral abdominal	2	1
Lower lateral abdominal	2	1
Cremasteric	1	1

(1 = slight; 2 = active; 3 = very active; 4 = with clonus.)

A questionable Babinski was observed on the left side; a questionable Chaddock and Oppenheim on both sides. Gordon and Schäfer were absent.

Myasthenic Status.—Flexion of the thighs was weak on both sides, more pronounced on the right. Extension of the legs was weak, more so on the right. Plantar flexion of the right foot was weak as was also dorsal extension of this foot.

Abnormal Involuntary Movements.—The patient had no tremors, twitchings, spasms or other involuntary movements.

Muscle Status.—All the muscles of the right leg were markedly hypertonic and the muscles of the left leg slightly hypertonic. There was slight contraction in the right hamstring muscles and the right calf muscles.

Abnormal Associated Movements.—The trunk-thigh sign of Babinski was present in both legs. The tibialis of Strümpell was present on the right. No other abnormal associated movements were observed.

GENERAL SENSORY. *Touch.*—Tactile sense was lost in a zone extending down from the level of the fifth lumbar vertebra including the left side of the penis and scrotum. Topognosis over this area was poor; there was no paresthesia.

Pain.—There was numbness in both legs evenly distributed; i.e., no patches but a gradual increase from the hips downward, more in the right leg than in the left. There was no other form of dysesthesia except a sensation of disquiet in both legs so that he moved them constantly, especially the right one. There was pain over the lumbosacral spine in a zone four inches long including the area over the fifth lumbar and first sacral. This pain, which was constant and dull, was worse at night, was relieved by rest and increased by exercise. The following pain responses were obtained from the galvanic current:

4 inches above umbilicus required	4.5 ma. to elicit pain
3 inches above umbilicus required	3.6 ma. to elicit pain
Line of umbilicus required	5.0 ma. to elicit pain
Below umbilicus required	6.2 ma. to elicit pain
Right thigh required	6.0 ma. to elicit pain
Back below T 12 vertebra	5.0 ma. to elicit pain
Inner surface right thigh	6.0 ma. to elicit pain
Back at T 12 vetebra	7.2 ma. to elicit pain
Back above T 12 vetebra	4.0 ma. to elicit pain

Temperature Sense.—This was greatly diminished over the area noted above and in places was completely lost. Heat sensations were normal over a V-shaped area on the back of the right thigh and leg and on the right side of the scrotum. Temperature sense seemed definitely more disturbed than touch or pain.

Vibratory, pressure and muscle-tendon sensibility were all normal. There was no disturbance in posture, weight or form sensibility.

Myotatic irritability was slight over leg and thigh muscles.

Electrical Reactions.—Faradic nerve excitability: There was more reaction from the same amount of current over the anterior crural and anterior tibial nerves of both legs than from stimulation of the musculocutaneous nerve of the arms. More galvanic current was needed to produce contraction in the leg muscles than in the arms. There was no reaction of degeneration.

Cranial Nerves and Head.—The cranial nerves were all normal including the special sensory, oculomotor and deglutitional innervations.

Mental Status.—The mental status was normal.

SYSTEMIC. *Cardiovascular.*— The pulse was regular and equal with no increased tension. Blood pressure: S 120, D 62. Heart was normal.

Rectal, Vesical and Sexual Control.—There was slight difficulty in beginning urination, otherwise normal; no changes.

LABORATORY TESTS. *Spinal Fluid.*—Appearance: clear yellow. Globulin: ++++. Cells: Red cells only. Wassermann negative. Blood count normal. Blood Wassermann negative. Urine analysis: Glucose present on normal diet. Sugar-free on diet without bread and sugar.

Summary.—The neurological findings showed spastic diplegia, anesthesia of the lower extremities and the lower part of the back and abdomen.

Diagnosis.—By means of faradic stimulation, Dr. Tilney found an area of diminished sensibility to fine touch at a level of the twelfth thoracic spinous process behind and coming around two inches above the umbilicus. He put the level of the mass at the ninth and tenth thoracic segments.

On *June 9, 1917*, the patient was operated upon by Dr. Adrian V. S. Lambert,[1] to whom we are indebted for the foregoing history. A laminectomy was performed. Dr. Lambert described the operation as follows:

"A median incision was made down to the spines of the vertebrae and the muscles were separated from the spinous processes and laminae. The laminae and spinous processes were removed by rongeur forceps. The dura was opened and a diffuse tumor was found extending for the entire distance of the opening and reaching above the opening and below it. The tumor was purple in appearance and irregular in shape, appearing larger in some portions than in others. It was very vascular and completely surrounded the nerve roots. It appeared to be in great measure behind the cord. Several pieces were removed for examination, with considerable hemorrhage. The dura was left open. The muscles were sutured over the tumor and open dural sac."

Pathological Report.—The specimen of the tumor consisted of a flattened mass of tissue about 23 cm. in diameter, which was blood-stained and friable and seemed to be composed of pinkish lymph glandular-like tissue through which were innumerable tiny bloodstained areas.

On microscopic examination the tumor was found to be made of a large number of small blood-vessels, the walls of which were thickened by a hyaline degeneration. Surrounding these vessels and forming the interstitial tissue between them were masses of round mononuclear cells with a deep staining nucleus about the size of a large lymphocyte apparently of mesoblastic origin.

The tumor resembled the one formerly removed except for the hyaline degeneration of the vessel wall. It might be considered either as an angiosarcoma or perithelioma, but was given the diagnosis of endothelioma.

June 23, 1917: The wound on the back had healed completely. The patient was able to move his legs much better than before operation, but they were still weak. He complained of tingling in the legs, but no persistent clonus as before. Ankle clonus could be brought out, but before operation the simple voluntary act of raising the leg brought on a clonus, which disappeared after operation. The patient was free from pain and quite comfortable. Beginning urination was a little retarded, but no incontinence existed. Defecation was normal. The patient was not aware of anesthesia, but believed his lower extremities were much less hypesthetic than before. The urine no longer showed the presence of sugar and carbohydrates were being steadily increased.

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[1] *Neurol. Bull.*, January, 1918, i, 1.

July 24, 1917: Hypesthesia was still present below the original line but very much less marked than before the operation. The left side was almost clear. Sensation was restored on the penis, but still absent over the left side of the scrotum. There was marked improvement in sensation on legs and feet. The hypalgesia corresponded to the hypesthesia. Temperature sense was still the most disturbed, being generally diminished over the whole of the right lower extremity and left leg. Sensation on the feet seemed to have cleared up more quickly than on the legs or thighs.

There was no spasticity, no permanent clonus, no Kernig, no Babinski or modification thereof; the deep reflexes were very active. The patient gained steadily in strength until he could support his weight on his legs and walk with the aid of a chair. There was no clonus and no anesthesia, and the general condition was good. In August he left the Hospital for his home in Tennessee. Quoting further from Dr. Lambert's report:

"From the records of the first operation it was evident that all the growth was not removed at that time. The improvement was slow but not unduly so for such a complete degree of pressure. The remaining growth undoubtedly increased slowly up and down the canal but gave rise to no symptoms for four years. It is possible that it continued stationary for some time after the operation and then suddenly took on a rapid growth. This does not seem probable, however. We believe that the tardiness in the appearance of the symptoms may be explained by the soft nature of the growth and its vascular character.

"At the last operation we found that the level of maximum pressure was not at the upper limit of the growth, in fact was well down below the level. The tenderness which was present over the spinous process at the site of the greatest pressure is comparable to that which is seen from time to time in cases of spinal cord tumors and is best explained by involvement or pressure on the dorsal branch which supplies dura and periosteum of bone at this level. It did not invade the nerve tissue.

"The complete removal of the tumor was quite impossible. Bleeding was most profuse on every attempt, and we contented ourselves with the removal of a portion for histological examination. The dura was left open to afford room for growth without exerting pressure on the cord. It was thought that a real improvement, which would exist over a long period, might be obtained in this way, as the tumor was slow-growing, having slight powers for compression and no tendency to invade the nervous tissue.

"The course of the patient's case subsequent to leaving the hospital has amply justified such a procedure, as his improvement has been continuous, and he now (December 1917) has completely regained the power of his legs. Endotheliomata vary in their malignancy, but this one certainly has shown no tendency to metastasis and has given rise to no cachexia. It is much too soon to give any accurate prognosis, but it would seem as though the ultimate outlook were bad."

The patient improved for a year after the operation just described, being able to walk fairly well with slight right leg limp. His knee jerks became practically normal and he was free from pain. He still had considerable weakness and some dullness of sensation. This condition remained stationary for a year, when he began to have symptoms and grew continuously worse until his third admission to the hospital, September 21, 1920. His condition was then about the same as before the operation three years previously. On this date a complete physical examination was made with the following findings:

General Appearance.—The patient, now

thirty-three years of age, was well nourished and well developed, and did not appear acutely or chronically ill. He lay quietly in bed, alert and responsive. Jaundice, cyanosis and dyspnea were absent. He was bald but had a normal distribution of hair on the body.

Head.—There were no scars or tenderness. The jaw was deviated somewhat to the right, with the right corner of the mouth higher than the left.

Eyes.—The conjunctivae and sclerae were clear. The pupils were equal, regular and reacted to light and on accommodation. The eyeballs were normal in position and movements. Vision was relatively normal. Eyegrounds: The discs were especially indistinct on the nasal side, and the right disc more

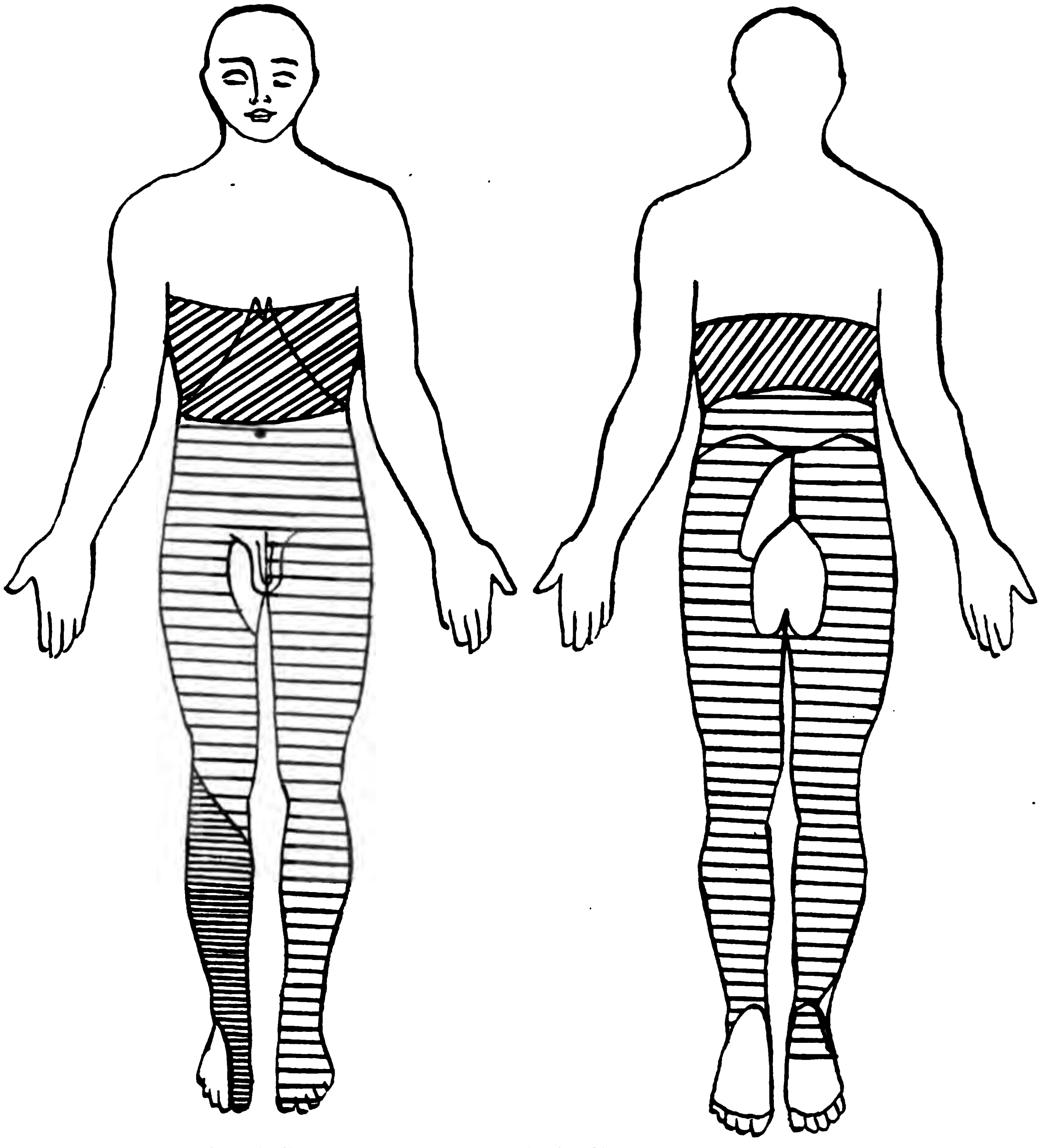

FIG. 1. Diagonal lines indicate region of hyperesthesia. Horizontal lines indicate region of diminished sensibility to touch.

indistinct than the left on the temporal side. The vessels were slightly tortuous and the veins turgid.

Ears were essentially normal.

the tongue not coated; and the teeth in fairly good condition. The tonsils were of moderate size, but apparently normal. The pharynx was slightly injected but showed no exudate.

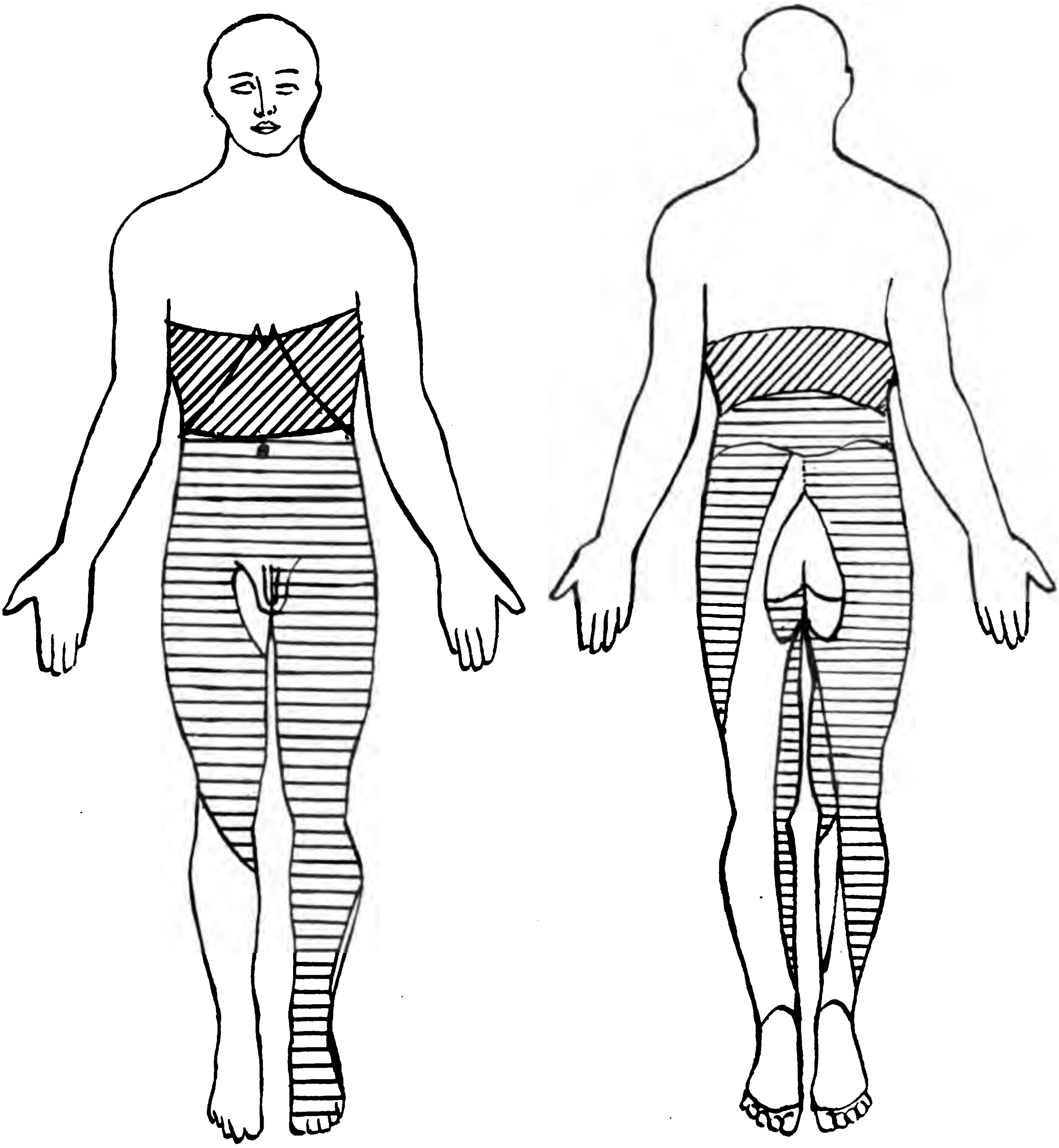

FIG. 2. Diagonal lines indicate region of hyperesthesia. Horizontal lines indicate area of diminished sensibility to pain.

Nose.—The breathing was better with the left nostril than with the right, the septum being deviated slightly to the right. There was no sinus tenderness.

Mouth.—The lips were of good color;

Neck was of good contour. There were no abnormal pulsations and the thyroid was not felt.

Lymph glands not palpable.

Spine.—There was some scoliosis. At the junction of the upper and middle third of the lower scar from the former operation, there was slight tenderness.

Thorax was well proportioned and symmetrical. No dilated veins were present and the mobility was apparently normal.

Lungs.—The breath sounds and fremitus were normal. There were no adventitious sounds and no cough. The chest wall was thick.

Heart.—No impulse or movements of precordium were seen. Apex beat was not felt but percussed 11 cm. to the left of the midsternal line in the fifth space. The heart was apparently not enlarged. There were no thrills and the sounds were of good quality. The aortic second sound was greater than the pulmonic second. The pulse was normal. Blood pressure 110-78.

Abdomen.—Contour was good, with a fair amount of subcutaneous fat. There were no abnormal movements or dilated veins. The right iliac crest was higher than the left. Palpation apparently normal, consistency rather resistant. No tenderness or masses were felt, nor were the liver, kidneys or spleen palpable. On percussion there was no distension, dullness or tympany.

NEUROLOGICAL EXAMINATION

October 4, 1920

Gait.—The patient was unable to walk without holding on to something for support or using a cane. The gait was both spastic and ataxic. He rather favored the left foot and lifted it higher from the floor than the right one.

Coordination.— Equilibratory: Standing with the feet together and eyes open, there was marked swaying. This was greatly increased when the eyes were closed. He was unable to stand on either foot alone with the eyes either open or closed.

Non-equilibratory: Finger-to-nose and pointing and pass-pointing tests were carried out accurately with either hand. Heel-to-knee tests were performed poorly on each side, the heel being placed above, below or to the inner side of the knee. If the heel were placed on the knee, he was unable to push it downward along the shin without having it slip off to one side or the other. There was no dysmetria of the upper extremities and no adiadochokinesis.

Skilled Acts.—There was no dyspraxia or speech disorder. Handwriting was normal.

Reflexes

DEEP

	Right	*Left*
Jaw	2	2
Pectoral	2	2
Biceps	2	2
Triceps	2	2
Radial	2	2
Ulnar	2	2
Wrist	1	1
Patellar	3	3
Suprapatellar	2	2
Periosteal uncrossed	Present	Absent
Periosteal crossed	Absent	Absent
Plantar	3	3
Achilles	3	4
Ankle clonus	Absent	Present

SUPERFICIAL

Epigastric	Present	Present
Upper lateral abdominal	Present	Present, but easily exhausted
Lower lateral abdominal	Absent	Absent
Suprapubic	Absent	Absent
Cremasteric	Absent	Absent
Plantar flexion	Present	Absent
Babinski	Absent	Present
Chaddock	Absent	Absent
Oppenheim	Absent	Absent
Gordon	Absent	Absent
Schäfer	Absent	Absent

Abnormal Involuntary Movements.—There were constant twitchings in the muscles of the upper and inner portions of each thigh, more marked on the right side. These were not of enough force to move the limbs.

Muscle Strength.—The patient was unable to rise from the recumbent position to the sitting without assistance of his arms. Opposed movements of the neck, shoulder, arm, forearm, hand and fingers showed normal strength on the two sides. There was moderate weakness in all movements of the thighs, legs, feet and toes. Plantar extension of the feet was not so weak as were the other movements. All other movements of the right leg were weaker than the corresponding ones on the left.

Muscle Status.—Volume: There was slight diminution in size of the muscles of the right calf and possibly, to a slight extent, of the lower part of the right thigh. The consistency and tone seemed normal.

Abnormal Associated Movements.— Not tested.

Nerve Status.—No observation.

GENERAL SENSORY.—There was a band of hyperesthesia to touch and pain extending completely around the trunk from the level of the tip of the xiphoid to slightly above the umbilicus. Below this, sensation was distinctly altered. Touch was but slightly felt on the anterior portion of the right thigh, though there was an area on the inner side of the right groin, the right side of the scrotum and the penis where it was distinctly felt. It was almost absent over the right leg except over the dorsum and upper portion of the foot. On the left side it was scarcely felt over the entire anterior portion of the thigh, leg and foot, and when it was perceived it "felt irritable" and not like normal touch. Posteriorly, cotton was not felt over the posterior portion of either thigh or leg, except on the soles of the feet and the external border of the right foot. There was also an area over the sacrum and buttock on the right side, and a smaller area over the lower portion of the buttock on the left side, where cotton was distinctly felt. There was anesthesia to pain below the umbilicus, down the anterior and inner portion of the right thigh to a little below the knee, except for a small area in the groin and on the right half of the scrotum and penis. Analgesia was also present on the left side throughout the anterior and inner portions of the thigh and leg. Posteriorly there was analgesia in the lumbar region and over the outer portion of the right thigh, and on the left side in the lumbar region and posterior portion of the thigh except for the inner portion of the buttock and down the posterior aspect of the leg to a little above the heel, and the inner portion of the sole of the foot including the great toe.

Temperature discrimination was impaired over all the areas where sensation was involved. Heat was practically never recognized. Extreme cold was sometimes noted but without regularity. Vibratory sense was absent over the feet, legs, pelvis, the lower three ribs on the right side and the lower two on the left. Firm pressure was usually felt as touch, but was no more painful in the analgesic areas. Muscle tendon sense: He was absolutely unable to tell the position of his toes or feet or to recognize rotation or slight flexion of the thighs. Flexion of the knees to a slight extent was sometimes noted. Compass test: He was unable to distinguish two points 12 cm. distant. Stereognosis: He was able to recognize by palpation the size, form and texture of any object placed in either hand.

CRANIAL NERVES AND HEAD. *Olfactory Nerve and Nose.*—Smell was normal.

Optic and Oculomotor Apparatus.—Vision, roughly tested, appeared normal. Eye-grounds: There was moderate swelling of both discs with obscuration of their margins. The veins appeared distended and tortuous. The pupils were equal, in mid-dilatation, and reacted normally to light and on accommodation. The palpebral fissures were equal. There was no exophthalmos, enophthalmos, ptosis or strabismus. The conjugated eye movements were normal. Nystagmus was absent.

Acoustic Nerve and Ear.—Hearing was good in both ears.

Trigeminus Nerve and Mouth.— The movements of the jaw were equal on the two sides. The corneal reflexes were present.

Facial Nerve and Face.—There was no evident facial weakness.

Glossopharyngeus and Vagus Nerves, Pharynx and Larynx.—The uvula was in the midline. The palatal and pharyngeal reflexes were present.

Spinal accessory nerve was normal.

Hypoglossus Nerve and Tongue.—Normal.

Summary.—The neurological findings indicated a lesion of the cord at about the level of the ninth dorsal segment.

LABORATORY EXAMINATIONS

Blood Sugar. September 24, 1920: 1.02 gm. per liter.

X-ray Examination of Teeth. September 24, 1920: Upper left first and second molars showed incomplete root coral work; upper right bicuspid, incomplete root coral fillings. There was a small area of rarefaction at the apex of the first bicuspid.

Urine Examination. September 24, 1920: Color amber. Specific gracity 1.039. Reaction acid. Turbidity none. Precipitate none. Albumin none. Glucose + + +. Acetone + +. Diacetic none.

Microscopic Examination: a few white blood cells.

Quantitative Analysis: 0.4 sugar.

X-Ray Examination of Skull and Spine. September 29, 1920: Irregularity of 5th lumbar transverse process which suggested trauma. There were six lumbar vertebrae. There was no indication of injury to the skull. The external occipital protuberance was prominent.

Blood sugar. October 7, 1920: .96 gm. per liter.

Urine. October 7, 1920: Sugar free; no acetone or diacetic.

Eye Examination. September 29, 1920 (Dr. Schoenberg). Bilateral choked disc. Swelling, right 4 diopters, left 3 diopters. There were numerous small round yellowish spots irregularly scattered in the right eye, suggesting also a star-shaped arrangement in the left eye. These spots are frequently found in diabetes, mild type.

October 9, 1920: Laminectomy performed by Dr. Lambert. A long, closely adherent growth was found reaching from about the sixth cervical to below the twelfth dorsal segments. As it was quite impossible to remove the entire mass, only the largest accumulations were removed. The patient left the operating room in a state of collapse, and died the following day.

AUTOPSY FINDINGS
(Dr. Pappenheimer)

Brain.—The calvarium was normal. The dura showed no increased tenseness. The longitudinal sinus contained soft clotted blood. The cortical convolutions were broad and perhaps slightly flattened. The superficial veins were engorged. The vessels at the base were delicate. There was no superficial deformity.

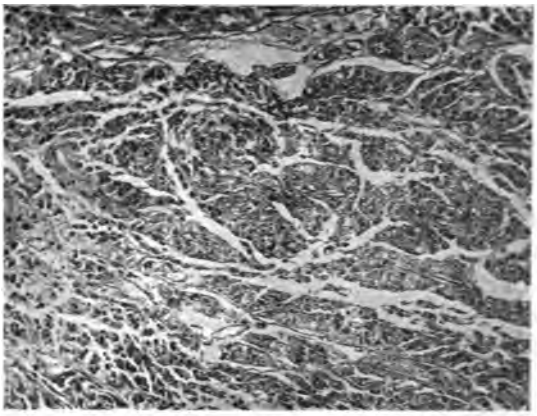

FIG. 3. Section of tumor.

The *middle ears* and the *sphenoidal sinuses* were normal.

Spinal Cord.—The cord was imbedded in an irregular tumor mass which was most abundantly present in the sacral region and about the filum terminale, but extended upwards as far as the upper dorsal region. The tumor tissue had a spongy texture but was not soft or friable. The color varied from dark red, resembling blood-clot, to grayish or yellowish white. The cord in places was greatly distorted by the tumor, but close inspection failed to show any invasion of the medullary tissue itself at any level. In the lumbar region, however, there was at one level an extensive necrosis of the cord appearing as opaque grayish softened tissue and involving all but a narrow shell of cord tissue. The spinal nerves passed through the tissue in many places, but there was no extension of the growth outside the vertebral canal.

Microscopic. Spinal Cord.—The tumor varied in its morphology in different situations, but in general presented a papillary structure. The center of the papilla was usually occupied by an endothelial-lined blood channel which was surrounded by a broad band of hyaline material staining pinkish red with Mallory's phosphotungstic hematoxylin, pink with Van Gieson, bluish with Mallory's anilin blue-orange G-fuchsin stain. There were no elastic fibers present in this stroma.

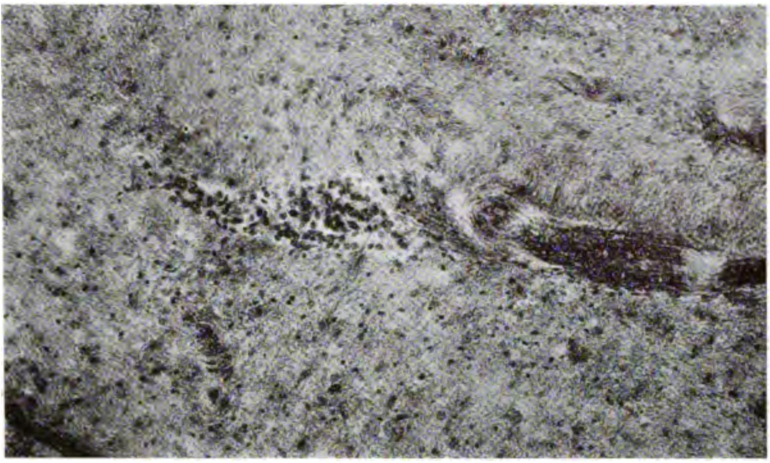

FIG. 4. Mid-sacral region. Extension of growth along small vessel into lateral columns of cord.

The cells composing the parenchyma of the tumor were ranged upon this fibrovascular papillary framework. Usually there were several rows more or less concentrically disposed, but occasionally the basal layer was in columnar rows, the long axis at right angles to the surface. The cells were usually elongated and of irregular shape. Their nuclei were large, oval, and stained deeply in the better preserved portions of the tissue. The cytoplasm was not sharply defined, often tapering into fibrillary processses. Definite fibrils, wavy, refractile and rather rigid in appearance, were formed on the surface of the cytoplasm and sometimes free between the cells. These were probably fibroglia fibrils since they stained bluish with phosphotungstic hematoxylin. Mitotic figures were very infrequent.

Blocks taken from various levels of the cord showed the following general relations:

4th Cervical.—One section showed a small mass of tumor tissue on the ventrolateral aspect, limited to the meninges and causing no compression or deformity of the cord. Pal-Weigert sections which did not include the membrane showed complete degeneration of the columns of Goll and a partial degeneration of the tractus cerebrospinalis dorsalis (direct cerebellar tract) on one side.

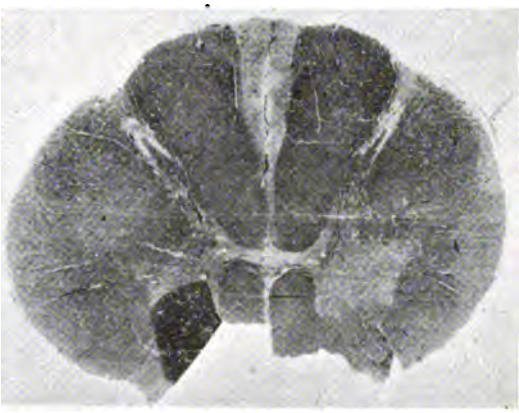

FIG. 5. Section at 4th cervical level. Pal-Weigert stain. Degeneration in posterior and lateral columns.

7th Cervical.—(Iron-hematoxylin, Van Gieson.) There was a block of tumor tissue lying external to the pia on the dorsal and lateral aspect of the cord. There was no invasion or deformity of the cord tissue. The tumor in this region was less cellular and showed abundant stroma. The pia was definitely thickened in the region of the tumor growth.

Mid-Dorsal.—The tumor formed a large crescentic disc between the pia and dura on the left(?) side, flattening the adjacent cord tissue but not invading it. The nerves and blood vessels were imbedded in the tumor tissue but did not appear to be compressed by it. There was an acute inflammatory exudate on the external surface of dura which showed fibrous thickening. The tumor was very cellular.

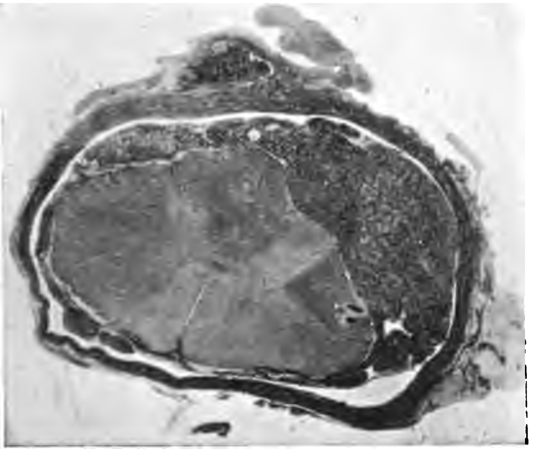

FIG. 6. Mid-dorsal region. Hematoxylin and eosin stain. Tumor compressing cord.

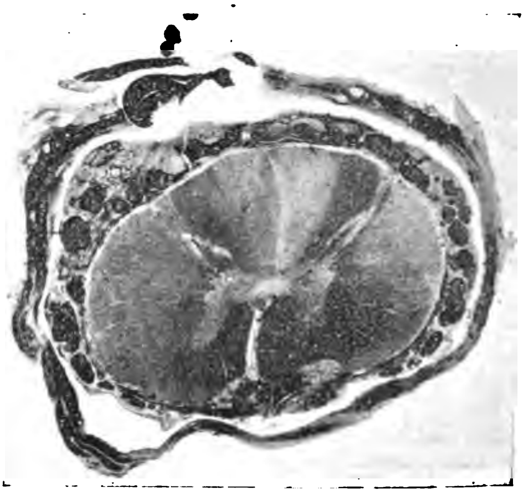

FIG. 7. Lower dorsal region. Pal-Weigert stain. Degeneration in posterior and lateral columns.

Lower Dorsal.—(Pal-Weigert.) The cord was practically surrounded by tumor tissue which filled the spaces between the nerve roots and blood vessels and extended into the ventral fissure; the anterior columns on one side were indented by the growth but the cord was not invaded. Degeneration (incomplete) was seen in the columns of Goll and in the lateral pyramidal and direct cerebellar tracts on one side. Included in the section, lying internal to dura, was a well stained parasite suggesting a larval crustacean. There was no obvious explanation for its presence in this situation.

Upper Lumbar.—There were large lobulated masses of tumor tissue on the ventrolateral aspects of the cord producing considerable distortion. The tumor tissue penetrated the dura with the nerve roots growing along the perineurium. Pal-Weigert section showed degeneration in the columns of Goll and in the crossed pyramidal tract on one side.

Mid-sacral Region.—The tumor tissue equaled the cord in bulk and had caused great distortion and deformity. It penetrated the dura in several places and in a few areas had grown through the pia and extended a short distance along the small vessels into the substance of the cord. A second block through the sacral region showed the cord displaced laterally and hollowed out into a thin cap by the large tumor mass. The pial limitation was, however, intact, and the nervous tissue was nowhere invaded.

Final Note.—An extensive tumor growth probably originating in the membrane of the cord and recurring after two operations. Following the classification of Mallory, the growth may be termed an "arachnoid fibroblastoma," and would generally be considered as an endothelioma.

(The remainder of the autopsy findings were excluded as irrelevant.)

CONCLUSIONS

There are several unusual features in this case:

1. Almost complete recovery from compression myelitis on two occasions when the portion of the tumor causing the pressure was removed by operation, the symptoms of compression occurring for the second time eight years after the first operation.
2. The unusual dimensions of this tumor, the relief of pressure allowing it to grow until it extended along the entire dorsal, lumbar and sacral portions of the cord.
3. The presence of choked disc. This is very difficult to understand and must have some explanation other than the mechanical theory by which we are accustomed to explain the occurrence of choked disc in cerebral neoplasms. A tumor of this nature would undoubtedly obstruct the free circulation of spinal fluid in the spinal subarachnoid space, but certainly no more so than tumors which compress the cord more completely than did this one. There was no indication of increased intracranial pressure on lumbar puncture. I have recently reviewed 86 cases of cord tumor to ascertain if choked disc had been observed before, and found it reported in but one instance, though it is possible that it was present in other cases and not observed, as no record of the eye-grounds was given. This one case reported by Fischer[2] was in a girl three years of age in whom at autopsy an enormous gliosarcoma was found involving different levels of the cord as well as the dura and vertebrae, but not involving the brain or skull. In our case, the brain and cranial cavity were carefully searched for evidence of tumor or anything which might have caused increased intracranial pressure or internal hydrocephalus, but nothing was found which could possibly have explained the presence of choked discs.

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[2] FISCHER. *Ztschr. f. Heilk.*, xxii.

DEFECTIVE DEVELOPMENT IN THE CEREBRO-CEREBELLAR CONNECTIONS

BY JOSHUA ROSETT, M.D.

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NEW YORK CITY

The patient, a deformed and apparently very feeble-minded Italian boy of thirteen, was brought to the Neurological Division of the Vanderbilt Clinic by his father, who wished to obtain a physician's statement that would help him to place the child in an institution.

The cause of the boy's deformities was the familiar one of injury at birth. After prolonged labor, terminated by instrumental delivery, he was born cyanosed and asphyxiated. Deep scars over both sides of the temporo-parietal scalp and the left ear, testify to the pressure exerted by the delivering forceps.

He was breast-fed. Although his general health during the period of infancy and childhood was very poor, his development, both physical and mental, was not so tardy as might be surmised from his present condition. He walked at two years and talked at three. The single abnormality in his development was the appearance of the teeth at the age of three.

The patient's hard-working, very poor and very ignorant parents, who had four healthy and well-formed children to bring up, completely neglected the education of their deformed child. They have not attempted to place him in an institution before now, because the boy has given them but little trouble. He has been docile and obedient. In spite of his infirmities he is as cleanly about his person as are other children of his social class, and he has never manifested any epileptic symptoms.

The appearance of the boy is striking. He is of medium stature. His well-shaped, strong limbs and body stand out in marked contrast to his deformities. He has a double internal strabismus, a stupid expression of the face, which is noticeably flattened on the left side, a misshapen nose, a scarred left ear, a scarred scalp. A drooping head, a stooping back, semi-flexed and abducted elbows, flexed wrists and widely spread out lower limbs, a good deal flexed at the knees and thighs, complete the picture of a thorough cripple. (Fig. 1.)

As he walks with a stiff, awkward, balancing gait, both his upper and lower extremities may be observed to vibrate with every impact of his feet against the floor. With a little support he can walk both on his toes and heels, but he staggers and falls upon any attempt to stand or walk on a reduced base unsupported, both with his eyes open and closed.

His non-equilibratory coordination is as defective as his equilibratory coordination. He can touch his nose, he can apply each of his fingers to his thumb, or touch the observer's fingers, but his fingers arrive at their aim only in the midst of wide oscillations of the entire upper extremity. When his elbow is supported, the oscillations are diminished and his hands and fingers are less hampered in the performance of a voluntary act. And the same is true of his lower limbs.

With respect to his intention-dyssynergia, the following point should be noted, that in carrying out a voluntary act, the muscles which move the particular segment or segments of the limb in the needed direction perform their function quite normally, and that the reason each respective distal segment has difficulty in arriving at its aim is because of the faulty fixation afforded it by each respective proximal segment. With the proper support to his elbow, he can move his hand and fingers in any given direction. The patient himself is conscious of the faulty functioning of the proximal segments of his limbs, and in order to enable his fingers to perform a skilled act he will grasp his forearm below the wrist with his other hand for the purpose of steadying it. His difficulty lies not in moving the distal segment of the limb in the needed direction and for the needed distance, but in the uncertainty of the base to which the distal segment is attached. Instead of being stationary, that base moves for an unexpected distance in an unexpected direction, and so upsets the patient's calculations. The following phenomenon, however, is seemingly inconsistent with the last statement: when the patient's scapula is adequately supported, and his upper limb performs a voluntary excursion which necessitates the simultaneous change of position of the limb as a whole and of its segments with relation to each other, the same oscillation of the limb is present as in an excursion which necessitates only the movement of the fingers and the fixation of the proximal segments of the limb. A closer observation of the patient's movements will explain this seeming contradiction, and the observation will be facilitated by the consideration of the following diagram. (Fig. 2.)

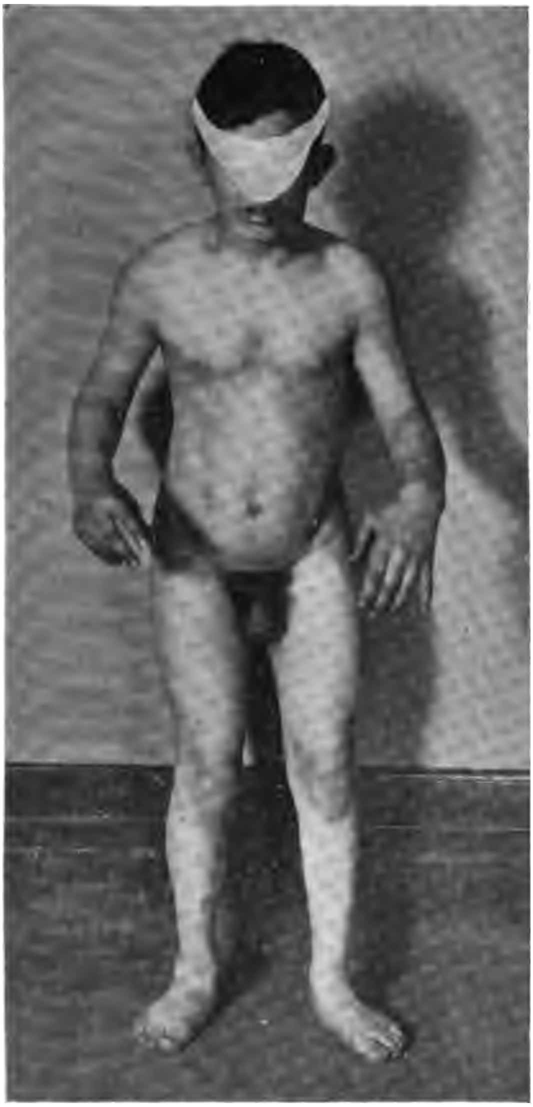

FIGURE 1.

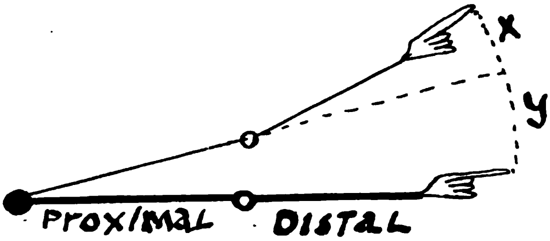

FIGURE 2.

In this diagram of the patient's upper limb, its two segments are severally and simultaneously in continuous motion until the distal extremity of the distal segment has attained the indicated object. The extent of movement of the distal upon the proximal segment is represented by the line X, while the total distance traversed by the distal segment is the longer line X + Y. The line Y, which represents the angle traversed by the proximal segment of the limb, represents at the same time a distance through which the distal segment was carried in a fixed position upon the proximal segment. In other words, the motion of the distal segment from its original horizontal position to the object above, although continuous, is composed of two factors—one, its own independent or active movement for a distance represented by X; the other, a passive movement, the extent of which is represented by Y. The muscles attached to the two segments served, during their contraction, a double function:

they moved the distal segment upon the proximal one for the distance X, and they immobilized the distal segment while it was carried through the distance Y. In short, the excursion of the index finger through the space $X + Y$ contains a factor of fixation; and muscular fixation being faulty in this patient, we must subtract this factor from any movement which might contain it, before we can decide whether the particular movement is normally or abnormally performed.

It will be observed that our patient, like all patients afflicted with muscle-dyssynergia, attempts to perform voluntary movements with undue haste. He can perform such movements better hastily than deliberately. Experience has taught him that the proximal segments of his limbs cannot be depended upon for any length of time as a basis for the movement of the respective distal segments, and so he is in haste to carry out the intended movements of the distal segments while movement is good. The muscular dyssynergia of this patient is not a dyssynergia of the muscles which move the distal segments of a limb in the needed direction, but one of the muscles which fix the respective proximal segments. His dysmetria is the fault not of the working machinery, but of the base upon which that machinery rests. His ataxia is the result of his astasia.

His speech is very defective in several distinct ways. His vocabulary, in both Italian and English, is even more limited than that of his parents. The paralysis of the sixth pair of cranial nerves, and the difficulty and awkwardness of the movements of his head, body and limbs, have resulted in a great narrowing of his visual, auditory and tactile experiences. Repeated experience with a multitude of objects and actions necessitates their naming, their classification and, finally, their symbolization. A reduction of such experiences must, therefore, be one of the prime factors of the poverty of language. In addition to this disadvantage the patient suffers from a high degree of dysarthria. The same dyssynergia with the resulting dysmetria which afflicts his body and limbs, hampers the movements of his organs of speech. Speech being difficult, it was natural that he should abstain from it, and his parents never took pains to encourage an exercise which, at its best, might serve only to annoy them. Yet, much as the nominal, the syntactic and semantic phases of the patient's speech have suffered quantitatively, neither of these phases has suffered qualitatively. The boy can name objects and actions, can form sentences and can understand the significance of objects, actions and symbols exactly to the extent of his experience with them. He can name a nickel, a dime, a penny, a pen, a handkerchief. He can repeat words and imitate movements. He answers to commands to stand up or to sit down. Moreover, he is capable of learning with a surprising degree of facility. With all the difficulty that he has in understanding English and in concentrating his gaze on the observer, he has been taught within a few minutes the meaning of "toe up" and "toe down," and to report with an astonishing degree of correctness the sensation of a tuning fork applied to various parts of the body. The injury to the brain which has affected the entire musculature of his body, including that of the speech organs, has spared both the formative and emissive phases of speech.

The boy's mental state cannot be characterized as one of feeble-mindedness until his potentialities for learning have been fully ascertained. A mere so-called "mental test" would be very misleading in his case. He is wanting in experience. He is uneducated in the widest sense of the word. If he does not know that five and five are ten, it is, in the first place, because his environment has been very largely devoid of mathematical conceptions—of almost any abstract conceptions, in fact. Yet he understands the meaning of good and bad, of more and less, and he is capable of evolving a number of the simpler relations. When asked to move his hand in a direction corresponding to that in which his toe was moved, he grasped the left wrist with the right hand (in order to do away with the fixation-dyssynergia of the proximal segments of his limb) and fulfilled the request in a manner which was but little less imperfect than that in which the average clinical patient fulfills a similar request.

The manifestation of his emotions is very misleading. His explosive, dull and hollow laughter, and the inarticulate whimper, would seem to stamp him as a hopeless idiot. The manner of both his laughing and crying are due to dysarthria. While the muscles of his diaphragm and thorax move rhythmically in the act of respiration, they function normally. But as soon as he attempts to fix the musculature of the thorax in definite positions to correspond with the alternating movements of the laryngeal muscles in the acts of laughing or crying, it fails him. The result is either an explosive, unmodulated sound or an inarticulate whimper. Add to this the fact that he cannot fix the musculature of expression for any length of time, as may be seen from the oscillation of these muscles upon his attempts at whistling, and the cause of the horrible manner of his laughing and crying becomes explicable on the basis of a faulty fixation of the successively proximal segments of the apparatuses of expression, speech and respiration.

The deep reflexes of the pectoralis, biceps and triceps, and the radial, the ulnal and the wrist reflexes, are normal on both sides. The suprapatellar is slightly exaggerated and the patellar considerably so on both sides. There is a tibio-adductor reflex from left to right, a transient clonus upon tapping the Achilles tendon, and a permanent ankle clonus.

The superficial reflexes are normal throughout, with the exception of the presence of the sign of Babinski in a very pronounced form. The dorsal flexion of the great toe cannot, however, be induced by any of the other methods.

The muscular strength is good throughout the body and extremities. There is a considerable degree of hypertonus in the lower extremities, and perhaps some degree of hypertonus in the upper extremities, neck and trunk as well. The hypertonus of the lower extremities becomes noticeably reduced after a few passive movements of the limbs.

The sensations of touch and pain, the vibratory and the muscle sense are normal.

Dr. Berens, who kindly examined the patient's eyes, reports that the optic, the third, fourth and sympathetic nerves are normal; that the abducens nerve is completely paralyzed on the right and partially so on the left; that the patient has a homonymous diplopia for 25 cm.; and that his vision is not bad. He does not assign any special importance to the small size of the palpebral fissures.

The condition of the motor part of the left fifth nerve is doubtful. On palpation the left masseter muscle appears to be weaker than the right. The weakness may be real, or the appearance may be due to the thinness and weakness of the overlying facial muscles. The left seventh cranial nerve must have been involved in the brain injury at birth. That the left side of the face had been paralyzed is proved by the observable flattening of that part. The boy's father informs me that food frequently accumulates between the teeth and cheek on the left side, which the patient digs out with his fingers. The occasional dribbling of saliva, which adds so much to the boy's feeble-minded appearance, is due to the same defect.

The left lung is thoroughly involved in a consolidating process, which is most probably of a tuberculous nature. Outside of this our systemic examination, including a blood Wassermann test, revealed nothing of importance.

General and Analytical Diagnosis.—The history of a prolonged and difficult labor is amply born out by the marks of the forceps, and places the patient's condition in the general class of birth palsies. The analytical diagnosis is more difficult. The most prominent symptoms of the patient are the following: A paralysis of the sixth pair of cranial nerves, a vestigial paralysis of the left seventh, a possible paresis of the left motor fifth, hypertonus, especially of the lower extremities, the sign of Babinski, and a fixation-dyssynergia of the entire somatic musculature. An injury to the complex made up of the sixth, the seventh, and the motor fifth, leads one to think of an injury to the tegmentum of the pons. The cause of the widespread muscular dyssynergia is somewhat more difficult to locate. Since the patient's voluntary muscular movements are intact, we may exclude, I think, the transverse fibers of the pons proper, where an extensive lesion must involve the pyramidal tracts. A primary injury to the entire cerebellum and to the tegmentum of the pons is not probable. Such an extensive primary injury would certainly involve the ponto-cerebellar angle, with consequent paralysis of the auditory nerves, and, above all, the occipital lobes of the cerebrum, which would be injured at least to the same extent as the structures which they cover. The patient would then be at least as blind as he is now astasic. A diffuse lesion in the tegmentum of the midbrain, beginning below the level of the nucleus of the fourth nerve and extending downwards along the midline into the tegmentum of the pons, would account for most of the patient's symptoms. Such an injury would involve the efferent tracts of the cerebellum above, and the nuclei of the sixth below. Midway between the two, the lesion might graze the motor nucleus of the fifth on one side or the other. The seventh nerve winds around the nucleus of the sixth, and would be implicated in a lesion of that nucleus. The sign of Babinski and the general spasticity of the musculature may or may not be due to a slight involvement of the pyramidal bundles by a diffuse lesion working its way from the tegmentum into the pons proper. The reconciliation of the two theoretically incompatible sets of symptoms—intact voluntary movement on the one hand and hypertonus and the sign of Babinski on the other hand—is fraught with certain difficulties in the present state of our knowledge. On the point of this incompatibility physiology is discreetly silent, clinical neurology vague, and pathology contradictory. The question becomes still more complicated in view of the fact that in an injury to cerebellar control as widespread as that in our patient, we are led to expect hypotonia rather than hypertonia.

The exact nature of the pathological process must, in this case, remain rather speculative for the present. I am inclined to the opinion that the injury was not the direct result of the application of the forceps with the production of meningeal hemorrhage. A meningeal hemorrhage that would reach, press upon and injure the tegmentum of the pons and midbrain to the degree that they were injured in the patient, must be extremely extensive, and would either kill the patient at once, or at least result in deep porencephaly, with consequent complete idiocy. I should rather favor the conjecture of a lesion in the encephalon as the result of prolonged uterine pressure upon the body of the fetus. Such pressure might cause serious edema or the rupture of a blood-vessel in the least supported parts of the brain, that is to say, the most central parts, among which must be included the tegmentum of the pons and midbrain.

DISCUSSION

Dr. Wechsler. I think this boy has all the signs of mental deficiency. He also has involvement of the pyramidal tract. I believe that the injury at the time of his birth resulted in an intracerebral hemorrhage and subsequently in a maldevelopment of the brain.

Dr. Polon. The boy undoubtedly shows some signs pointing to pituitary involvement—the large breasts, the enormous genitalia. I do not think the lesion should be placed altogether in the cortex. In view of what appears to be a marked predominance of his bulbar signs, or pseudobulbar signs, I believe the bulb may be considered as the seat of the lesion.

Dr. Osnato. It seems to me that this case can be explained on the basis of an injury to the cortex involving the opposite half of the hemisphere. We know that injury in one half of the cerebral hemisphere causes an atrophy in the other half; so that a double injury here would explain all the symptoms, the pyramidal tract signs and the asynergia. I do not believe that there is a pontine hemorrhage here, that is, in the pons itself, because I do not see how you could get pyramidal tract signs in that event without also getting signs of the fillet.

Several hemorrhages rather than a single hemorrhage are apt to occur with these birth injuries. I rather incline to the idea of a basal hemorrhage and a large double hemorrhage, probably extra-dural.

Dr. Strong. This case is, of course, in a general way one of the heterogeneous group styled infantile cerebral palsy, not, however, of the more usual type. It is certainly a very puzzling one to me.

It is not easy to give all the phenomena even an accurate clinical title. For example, the posture is a peculiar one. It is mainly a flexion posture and one which might be assumed by one suffering from an equilibratory ataxia. From this point of view one might be tempted to say it is because of this permanent defect since birth that he has assumed this constant, though not, as I understand it, fixed posture. Then, too, regarding the abnormal movements he makes, are they to be characterized as ataxic or are they more of the "choreo-athetoid" type, especially perhaps here choreiform abnormal involuntary movements? I would like to know what the opinion of the conference is as to this latter question of observation. The question as to whether the lesion involves the cerebellum itself depends partly upon the answer. Besides this difficulty and the complicated character of the case, there is also the lack of good cooperation, which makes one uncertain as to the exact sensory and even motor status.

Regarding the involvement of the pallium, one of the most decisive points is the question as to the boy's intelligence. I am considerably in sympathy with what Dr. Rosett says about the liability to underestimate the intelligence of one whose means of expression are and always have been so limited and who has not been afforded much opportunity to make the best of what he has. Nevertheless I do not think he can be accredited with his full share of intelligence on account of the reactions mentioned by Dr. Rosett inasmuch as these can be performed by a normal individual considerably younger than the patient. I was somewhat inclined at first to place the whole lesion in the cerebral hemispheres, attributing the instability of movement to the involvement of the pallio-pontile system. While perhaps this might explain the instability of standing, I doubt whether it would explain the other abnormalities of movement.

We might also be tempted to place part at least, of the lesion in the corpora-striata. The dysarthria, excessive laughter and crying and possibly the abnormal movements suggest this, superficially at least. On the other hand there is an absence of the more rigid athetoid character in the movements and the crying and laughter are appropriate though excessive. The latter are thus more like pseudo-bulbar phenomena. Then, too, there is the double Babinski, though I am not sure that the extension of the big toe when elicited simply as a part of more generalized movements of an athetoid character is to be interpreted as necessarily a sign of pyramidal involvement.

Accounting for the movements and possibly the pseudo-bulbar symptoms, if they be such, by a midbrain lesion or lesions appears to be negatived by the absence of third nerve (and probably absence of sensory) symptoms. A lesion in the pons proper (not the pontile tegmentum) would give such a marked pyramidal lesion that there would be a spastic paralysis obliterating the movements actually present.

Then, too, there is the abducens involvement which would appear to necessitate the assumption of some basal lesion (hemorrhage?) involving this nerve, in addition to the forebrain lesion. Nor does it seem certain to me, as stated at first, that there may not be a lesion of a purely cerebellar character.

Dr. Casamajor. Dr. Rosett seems to think that a mental test would show the child's mentality lower than it really is. I am inclined to think that this is not the case. The Binet tests, it is known, come very close to the mental age of the child in spite of education or lack of it. I think it would be well to have a Binet test on this child.

Dr. Polon. On the basis of a porencephaly it is difficult to explain the involvement of some of the nerves, such as the 5th and 6th. On the basis of a basal lesion it would be difficult to explain the involvement of the 7th and the mental defect. For this boy certainly has mental deficiency. He has the mentality of a child of four. I wondered if there was not evidence in favor of the lesion being a multiple one.

Dr. Strong. I would like to know what the opinion of the conference is about the movements that the boy makes. Are they cerebellar ataxic or choreiform movements?

Dr. Polon. Oppenheim says that paralysis is not necessarily an indispensable element of infantile palsy. It may be merely spasticity, spasticity that is not always manifest. It may manifest itself only in excitement. This boy's movements seem to me to be due to spasticity. When he attempts to carry out an act his muscles become rigid. He attempts to overcome the rigidity and the asynergia comes into play. On the other hand, if one limits the muscles that take part in the movements, the movements are better performed.

Oppenheim says that such brains as these show porencephaly. The amount of injury induced by the birth palsy may not necessarily be due to the forceps.

I consider the nature of these movements as being that essentially due to spasticity.

Dr. Casamajor. When one considers that the cerebrum does not fully develop until quite a while after birth, differential diagnosis in such a case becomes still more difficult. There is a continued development of the cerebrum for months after birth, and the question must arise as to whether this child could have been in this state when he had been normal at birth. From the small head one feels that there was a prenatal defect and that the whole thing cannot be a birth injury. But that is purely a matter of opinion.

As to the point Dr. Strong raises, whether this is ataxia or choreo-athetosis, I think before one can answer one has to be sure of the definition of the terms. I should not want to pass an opinion as to whether it is choreo-athetosis or ataxia. I think it is a little of both and a little of neither.

Dr. Tilney. I have seen this boy for a moment only. But from what I have seen of similar cases, it impresses me that we are dealing with choreo-athetoid type of movement; at the same time he certainly has a marked degree of asynergia, and I can see no logical reason for being averse to putting the lesion in more than one place. In the light of his birth injury, why are we restricted to any one locality of the brain? Is it not fair to presume that under the circumstances attending his birth a large territory may have been involved? It is not such a far cry from the striatal to the rubral center. The red nucleus acts more or less as a clearing house for both lines of transit.

I am inclined to think that there is a good deal of asynergic defect in the case.

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NEUROSES IN BUSINESS LIFE

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Modern social legislation has stressed a point—responsibility of employers for the health of their employees—that has made the psychoneuroses a problem of as much importance in industrial life as it has been to armies in war. The traumatic neuroses in the industries have presented a many-sided difficulty and one which to-day is far from its ideal solution. Workmen's compensation laws and compulsory accident insurance have been steps toward a solution, and while they have been steps toward justice for the cases of organic injury and disease, for the pscyhoneurosis they have been but stimuli for the development and persistence of sickness.

Our experience with the war neuroses increased our understanding of the mechanism of all neuroses. The true nature of these conditions was understood only when we had discarded all ideas of a traumatic causation, and looked underneath, at the personality of the patient, his psychological adaptations and methods of handling a dangerous and distasteful environment. The symptoms of the psychoneuroses developing in the army at home were not different from those of the troops in the line, nor was the psychological mechanism in the least different. The point where the neurosis occurred was determined in each case only by the ability of the individual to handle the environment in which he found himself, and to face the problem which lay next ahead of him. Some developed the neuroses before registering for the draft, others only after months in the trenches.

To study only those industrial neuroses in whose causation an element of trauma may be included and where compensation is a possibility, is to observe only one phase of the question. Were desire for compensation the only factor, the problem would be simple. It would then be a question, not of neurosis but of malingering; in other words, the desire to be sick would then be conscious rather than unconscious as it is in the neuroses. Certainly this is untrue in the vast majority of cases. Osnato stresses this point as follows: "The Compensation Law of this State (New York) allows a maximum weekly indemnity of $15. Certainly this sum can be earned by the average laborer even though he is not particularly skilled. Besides this, many patients are married and have families and there is every reason for them to return to work as soon as possible." Again, were these neuroses unknown in business life, where there can be no claim for compensation, there would be less difficulty in their interpretation. This also is far from the truth. Such neuroses are very common but usually pass unnoticed, for the monetary asset value of the neurosis is absent, the patient has no claim on his employer, and the relationship of the work to the neurosis is overlooked.

During the past two years the writer has had the opportunity of examining a number of young men and women with such neuroses in relation to their work. The neurosis was different in each case and quite unassociated in the patient's mind with the employment. However, the mechanism that underlies the compensation neurosis existed, and I feel sure that had any of these patients sustained a trauma connected with his work, he would have developed the typical traumatic neurosis, the condition most frequently studied.

In arranging this material, I have chosen a few cases and grouped them very loosely under three headings.

1. The inadequate;
2. The dissatisfied;
3. The generally maladjusted.

No case can be considered as belonging to but one class, for elements of all these appear in each. There is one outstanding feature in every case: no one was satisfied with his job. It is doubtful if any one of us is satisfied with his job and his life. While all these patients had many reasons for being satisfied with both their lot in life, and the remuneration and the type of work, yet there were many causes for dissatisfaction —reasons, usually, the patient was ashamed of or did not want to admit, even to himself. These latter reasons were more or less unconscious, and it is in this conflict of reasons that we see the causation of this neurosis as of every other neurosis, both in civil life and in war.

THE INADEQUATE

The inadequate is the individual who is not equal to his job. When he finds himself in a position which is beyond his ability, he becomes fearful that he will not succeed. He dislikes responsibility and would prefer to earn his living in an easier way. To do so, however, would reduce his income, and he finds himself with a real conflict on his hands. Primarily, he is afraid of failure, and his illness (neurosis) is an excuse for failure and a reason for not attempting success. The onset may be at any time and is usually induced by a change in the patient's working conditions, either a promotion to a position of greater responsibility and beyond his capacities, or a misfortune which complicates the business life by requiring greater responsibility and activity. Since fear lies at the basis of the mechanism, anxiety is the principal symptom of the clinical picture.

CASE I. Male, age thirty-six, married, was referred for mental examination because he was acting peculiarly. He was anxious and worried and felt unable to do his work. He had remained away from his place of employment for a week and could offer no adequate reason for having done so.

History.—The patient is an only son with two elder sisters, and his father had died when he was two years of age. He was the baby and pet of the family, being allowed always to have his own way and pretty generally spoiled. He did fairly well in college, interesting himself more in athletics than in his studies. After graduation he worked in different banks with regular advancement. In 1915, while serving as treasurer of a small country savings bank, he began to lose confidence in himself, disliked the responsibility, did not want to go to work and was especially afraid to write letters. This resulted in a "nervous breakdown," and on the advice of his physician he went South. After a few months in a sanitarium he recovered completely. He then obtained a clerical position in a bank and did well until eighteen months later when a similar condition recurred. This preceded a contemplated step which would increase his responsibilities to a considerable degree—his marriage. After two months' vacation he recovered, married and came to New York. He soon obtained a position as a traveling salesman for a large trust company. This work he found very congenial. His orders were definite, even his "selling talk" was written out for him, and under these conditions he was quite successful. His success became a bit too much for him, for it brought to him advancement and more important work to do. This meant more responsibility, the thing he had always avoided. His work was now described as being of two sorts— very efficient when on the road and most inefficient when doing work of an administrative sort in the office.

In August, 1919, a new responsibility was thrust upon him. His wife, nearing parturition, went to live in a distant city with her mother until the birth. He gave up his apartment and went to live in a boarding house. This existence he found most irksome. He worried about his lonesomeness, the coming increase in his family, and the increasing responsibilities of his position at the bank. He became fearful and felt that he could not do the things asked of him. His family and financial responsibilities, however, prevented his relinquishing a remunerative position as he had done before. In November, 1919, he was given his most important business assignment—to address a meeting of bankers in the West. He worried about it constantly and feared he could not trust his own judgment. He would not ask for help for he feared to endanger his position. Having completed his assignment he was certain he had made a complete failure of it and on his return to New York he was afraid to return to the bank and face his superiors. Hence he stayed in his room for a week before returning to work. During this time he felt sick, nervous and fearful. He could account for all his actions during this time and stated merely that he was "too sick and nervous" even to think of returning to the bank.

Case II. Male, age forty-three, married, was examined in February, 1921. At this time he had many complaints of a definite anxiety character. He had headaches, dizziness, tremors, bad dreams, insomnia, fear that he was going insane, that he had a lesion in his brain, that he had syphilis, and other fears of a less definite character. The idea of syphilis was so persistent that he had submitted voluntarily to a lumbar puncture to determine it. He was definitely disappointed at the negative result.

This man had been very successful as a jobber in textiles. He had married eleven years ago but had gotten little happiness from his married life. His wife—sexually frigid—had never shown him the sympathy he needed. All he got of love was at the cost of a struggle. There was no cordiality in the home and hence he put all his energies into his business. His business became his whole life and he reaped a considerable measure of success. During the war and after it, with the rising price of textiles, he soon found himself well off with affluence apparently just ahead of him.

In February, 1920, the market "broke." This meant heavy losses for him and he worked day and night to "juggle" his orders so as to save some of the loss. At length one of his largest customers threatened him with suit for delayed delivery. This was the situation which he was inadequate to meet. On questioning he admitted that the idea occurred to him, "If I only get sick perhaps they would take pity on me and not crowd me too hard. Then perhaps things might change and I could work my way out of it." Almost immediately his neurosis developed.

He complained of some of the symptoms mentioned and was advised to take a rest. With his wife, he went to a nearby seaside resort and one night he casually mentioned that he would like to commit suicide. At this his wife was thoroughly frightened and began to treat him kindly for the first time in years. His neurosis now rapidly became worse, for it acquired further value in the kindness and consideration it secured him from his wife. He made two or three half-hearted attempts at suicide but now admits that he never did them except when he was sure he would be rescued. While in a sanitarium with psychotic patients he picked up a number of new symptoms and much medical terminology. The threat of a suit was still hanging over him and his wife was beginning to lose patience with his disease. Syphilis was the last symptom he acquired and he stated,"If I haven't got syphilis I don't know what is the matter with me and I am doomed." On advice, the suit question was settled and his wife was instructed to improve her attitude and he is now going to his office every day.

THE DISSATISFIED

This group is the largest, for it includes the great majority of the neuroses of business life. We nearly all belong at least in the borderland of this group, but most of us do not develop neuroses to solve our difficulties. The neurosis arises in the individual who likes the remuneration he gets from his work but dislikes the work itself and the type of life it forces him to lead. The symptoms are not usually severe and most of these patients struggle on without applying for medical aid. However, should the patient suffer an accident for which the employer could be held responsible, a typical traumatic, litigation neurosis might easily appear. Many of these patients are young men who have served in the army and have had considerable difficulty in readjusting themselves to civil life. Life in the army, while not an ideal one, nevertheless is not totally unpleasant. It has its advantages in the strong patriotic emotion, a certain prestige, and a freedom from great financial worry which appeal even to the moderately indolent. These cases of mine might just as well be considered "Post Service Neuroses" as Neuroses in Business Life. As a rule these patients recover rapidly under analytic and suggestion treatment.

CASE III. Male, age twenty-five, single, was referred for examination on October 22, 1920. He complained of depression, sensation of pressure in the back of the head and general nervousness.

History.—This young man entered West Point Academy in 1916, and failed in 1918. Immediately he enlisted as a private in the army and was commissioned Second Lieutenant in the same year. He did not serve overseas and found the life so much to his liking that he was much disappointed when he was discharged in October, 1919. Following this he did nothing for three months, seeking to find himself in an environment he disliked. In January, 1920, he secured a position as clerk in a large bank. Although his work was amply efficient, he yet took no pleasure in it. He still longed for the army. An uncle, whom he much admired, was a Colonel and it had been his ambition and that of his mother that he be an army officer.

During the summer he suffered from restlessness and pressure in the back of the head and was given a month's vacation. On his return to work his attitude toward it had not changed but he stuck to it through the necessity of earning his living. In the early part of October the patient contracted a severe cold and the neurotic symptoms above described returned. He felt weak and fainted while at work and was taken home. He had not been able to work since and was still in doubt whether he would return to the bank or try to get into the army.

CASE IV. Male, age twenty-eight, single, was referred for examination December 11, 1920. He complained of cold sweating of his forehead and hands, nervous attacks, palpitation of the heart and pains in the back of his head, all of about one month's standing.

History.—This man was born and up to 1917 had spent his life on a plantation in a far southern state. He went into the army in 1917 on the draft, but did not care for it. In December, 1917, he contracted pneumonia and following a short leave at home he returned to his company. He did badly from now on and complained of his heart. In the early part of 1918 he was discharged from the army with a diagnosis of neurasthenia. This diagnosis did not please him and after his return home his cardiac palpitation persisted. The family physician diagnosed "hypertrophy of the heart" and his cardiac palpitations soon disappeared.

In October, 1918, he came up to New York to make his way in life. While he loved the ease of his plantation life yet he felt the opportunities too limited outside of a big city. At first he worked for a machinery company and in January, 1920, secured a clerical position with a trust company. The work was not difficult but very congenial, and the opportunities for advancement appealed to his ambition; but boarding-house existence and riding in crowded subways was most distasteful to his indolent, comfort-loving southern temperament. He hated New York and all New York meant to him, except the chances for wealth. He stated that if he had money he would not live in New York. His condition was similar to that of the war neurosis. New York was "the line" for him in which he was forced to stay by his ideals of ambition. His plantation home was "Blighty," a place of ease and comfort. In a man who had previously had neurotic symptoms directed toward his heart, it was not surprising that they should have again localized there when he found himself with a psychological conflict to solve. A further complication in his life may have accounted for some of the anxiety symptoms he showed (the sweating and nervous attacks). He had been engaged for the past eighteen months, but felt he could not marry on his present income. He spent most of his spare time with his fiancée, and the repressions he forced on his amorous instincts may have been an added etiological factor. Anxiety is the usual coloring of the "Engagement Neurosis."

Case V. Male, age twenty-eight, married, was examined May 7, 1920. He was suffering from nervousness, inability to concentrate, and morning nausea of three weeks' duration.

History.—This young man was one of those fine, clean-cut specimens of young American manhood who commanded our troops in France. He had served almost continuously in the trenches for eighteen months as an infantry officer in two National Guard divisions with most active service. He was twice promoted, coming out at the Armistice as Captain of Infantry. During this time he remained continuously with his troops, never having been sent back for any cause. After his discharge from the army he entered the employ of a business house and was considered a most valuable employee. In November, 1919, he married and his home life was ideally happy. After a few months he began to be dissatisfied with his work. He felt his opportunities were limited and that he was wasting his time. He wanted to give up his job and get another, but felt he could not do so with his wife to support. His position now paid him well and he was certain that if he should change, his salary, in starting with a new concern, would be lessened. With the high cost of living he felt this would be an unnecessary hardship for his wife. He began to lose interest in his work and felt that he was not doing well, although his superiors were amply satisfied. He feared he was not making good and this fear was the reason for his inability to concentrate. His mind was not on his work. Soon he became nauseated in the mornings, while riding in the subway on his way to his place of business. He would have to leave the car a station or two before his destination because he feared he would vomit. He never did, however. He blamed the air in the subway for his nausea and went downtown on the elevated road. The result, however, was the same; he became nauseated and had to leave the train and walk the rest of the way. He then tried going to work on the surface car, with exactly the same result. It was obvious that his nausea was occasioned not by riding in cars but by his dislike for his work and place of business. It is cheering to note that he never was nauseated when riding home in the more crowded subway at night. In talking over the matter with his superiors he discovered that his opportunities in the company were much greater than he had believed and his symptoms soon disappeared.

THE GENERALLY MALADJUSTED.

Possibly it is not proper to include this group in this paper, for the difficulties of these individuals in business life are only a part of their general maladjustment. The maladjustment to the home life is of much greater importance, and the work is a means of escape from the home as in the case I report. However, these cases deserve consideration, for they are the ones that after a slight trauma most easily become typical litigation neuroses.

Case VI. Female, age twenty-seven, single, was referred for examination on October 25, 1920. She complained of inability to do her work as she used to, poor appetite, disturbing dreams, trembling of the hands and knees, pain around her heart, fatigableness, excessive perspiration and "nervousness" (anxieties).

History.—This girl was the eldest child and only daughter of a family of four children. She had always been kept very close to the home and was apparently much attached to her rather domineering mother. She had few friends and no men friends, as her parents did not approve of the men she met. Her life at home was quite colorless and uninteresting. In November, 1919, she secured employment in a banking house. She was conscientious and capable and was given an excessive amount of work to do with insufficient help. Often she would have to sort and distribute 4,000 checks a day and remain at her work nine to twelve hours at a time. She did not complain of this, for she loved her work and took an enormous amount of pride and pleasure in it. It was her only interest and the only self-expression in her life. She became fatigued, and many nights after getting home late would fall asleep before her dinner was ready. She cut herself off from all entertainments and amusements.

Such a strain could not continue, and in July, 1920, she had a "nervous breakdown." She trembled violently when starting for work one morning and was put to bed. Following this her neurosis developed rapidly, the symptoms above enumerated appeared, and she was given a long vacation. In September she went to the country, but sprained her ankle after three days and had to return home. As her fatigue disappeared her neurotic symptoms persisted. This girl had learned but two methods of meeting her psychic problem. One was overwork and the other the neurosis. Neither was satisfactory and she was face to face with a real quandary. She decided to return to work, but when starting on the morning of October 18th, she became unconscious while standing up in the hallway of her apartment. She was stiff and rigid when put to bed, and she remained unconscious for one hour. She felt weak and "shot to pieces." The next day she had recovered. Following advice to work less, and play more away from her home, she has made a very good recovery.

The great amount of enthusiasm this young woman put into her work shows that her work was to her a substitute for some other interest she could not express, and, while it undoubtedly kept her mind off other things and was emotionally satisfactory, still the gradually increasing fatigue made it physically impossible for her to continue this mode of expression; hence the neurosis as the only means left for self-expression and getting away from her problems.

The similarity between the neuroses of business life and the war neuroses is quite obvious, and one wonders why they are not more common. Certainly, business life in a large city like New York is artificial enough, and it is surprising that the human animal, developed under such different conditions, is pyschologically able to endure it. Dissatisfaction is ever rife throughout industrial life and the neurosis is a way out of a difficult problem.

When an injury for which an employer can be held responsible occurs such dissatisfaction can be capitalized unconsciously and the traumatic neurosis becomes the logical sequence. Bailey has emphasized the other side of this same picture in describing his experiences in the wreck of a "Banker's Special." Of the 150 men on this train, all men of position and fairly well satisfied with their lives, "not a single one developed a traumatic neurosis and no claims of any importance were brought against the railroad company."

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CYST OF THE LEFT CEREBELLO-PONTINE ANGLE

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WITH REPORT OF OPERATION BY DR. ELSBERG AND SPECIAL REPORTS ON THE EYES BY DR. CONRAD BERENS AND ON THE ACOUSTIC NERVE BY DR. RANDAL HOYT

NEW YORK CITY

The syndrome of the cerebello-pontine angle is now so well known as to lend particular interest to its variants in which the symptom-complex is so incompletely developed as to invite lively diagnostic controversy. The following case is an instance of this nature:

The patient, M. G., female (No. 30635, admission No. 69521), was admitted to the Vanderbilt Clinic January 12, 1921. She is twenty-one years of age, American.

HISTORY. *Complaints.*—Thepatient complains of:

(*a*) An inability to see straight, this being a tendency to see two objects or possibly merely a blurring of images.

(*b*) Dizziness; everything goes up and down with some tendency to go to the right. This phenomenon is produced by turning the head to the right when looking at an object.

(*c*) An impairment of hearing on the left side.

(*d*) Unsteadiness in walking due to trouble with the left lower extremity.

(*e*) Weakness, tremor and unsteadiness of the left hand.

(*f*) Does not think as clearly as formerly, feels stupid and sleepy.

(*g*) Cannot chew well on the left side, cannot bite as well, and food gets lodged there.

(*h*) Several attacks of severe, lightning, burning pains in the left cheek near the nose.

(*i*) Deeper pains which appear to go through the head on the left side from forehead (supra-orbital region) to occiput.

(*j*) A feeling in the head as though the left eyebrow wanted to pull upward, making her wish to pull it down.

(*k*) Suboccipital pains.

(*l*) She feels cold and hot objects more in the left side of the mouth.

(*m*) A queer feeling in the left outer thigh. This was preceded by a creaking feeling with pain in this location. She also has a lame feeling in the left leg near the knee after walking.

(*n*) When she stoops and gets up there is some buzzing in the left ear; but ear noises do not appear to have been otherwise present. There is apparently some feeling of rush of blood to the head when stooping.

Further description in detail of the symptoms just enumerated:

(*a*) The inability to see straight was first noticed toward the middle or end of 1918. For example, when she went to church and looked at the minister he would appear to have two heads, one above the other, which would come together if she looked at him steadily long enough. Signs were blurred. She could see all right with one eye, either left or right, but there was an uncomfortable feeling around the left eye, so she closed that one. This trouble began gradually. She thinks it is affected by fatigue and is getting worse.

(*b*) The "dizziness" began probably toward the end of 1918 or early in 1919. As already stated, this appears to consist of a wavering of objects which seem to go up, together with a tendency to go to the right and get hazy. This sensation is produced by turning the head to the right when looking at an object. It is worse just after getting up and is made worse by anything which "puts her in bad condition," e.g., lack of sleep. If she has had a good rest it does not bother her. This dizziness is getting worse.

(*c*) The impairment of hearing on the left side was noticed first about July or August, 1919, and apparently came on with the rest of the earlier symptoms. It has grown worse.

(*d*) The unsteadiness in walking has been noticed since about July or August, 1919. It has bothered her especially in going downstairs. In walking she stubs the left toe. She thinks the left foot does not go straight on account of weakness. A lurching and unsteadiness of gait is attributed more to this than to the dizziness, and can be corrected by paying attention to it rather than by looking at it. This trouble has developed gradually and she thinks it is still getting a little worse.

(*e*) The trouble with the left hand has developed more rapidly than the trouble with the left foot. In 1917 she could play the piano, but the left hand was weaker. There was no "jumping" then. July, 1919, she first noticed that she could not play the piano at all with the left hand, the fingers would shoot off and hit the wrong keys. Two months ago, she especially remembers, she picked up a glass while talking to her mother, and, as if some one hit her hand, it flew out of her hand and was broken on the floor. The first indications of the trouble were thus apparently noticed three years ago in playing the piano, and consisted of a "weakness" of the left hand, which was not more easily fatigued but just weaker than formerly. The "jumping," which came later, is worse, but not the weakness.

(*f*) The stupid, sleepy feeling was noticed first in 1917, and was worst in May, 1919. She thinks this began with the rest of the earlier symptoms. She also thinks her memory is not quite as good as formerly; for example, she does not remember dates well. There has not been much change since May, 1919, but it is possibly somewhat worse.

(*g*) The weakness in chewing on the left side she thinks was first noticed in the summer of 1918. It is getting worse.

(*h*) The attacks of pain in the left cheek first occurred in May, 1919. They began suddenly and were like a red-hot shooting feeling, each lasting a second. They were very severe and occurred on an average twice a day for a period of about a week. In October, 1920, she had another attack. The pains were still more severe and she felt as though the face were drawn; tears came to the left eye. The pain lasted half a minute and she had two more, the last for about a minute. She also had other stabs while eating which would make her stop and cry out. These pains only occurred for about two days and she has had none since. She thinks possibly they may have been evoked by washing the face or eating; at least they came on then.

(*i*) The deeper pains from forehead to occiput are not severe. They were first noticed about June or July, 1919. They occur about once a week. They are not getting worse.

At the same time there have been some pains in the left back of the neck, not severe but more disagreeable. These date from 1918, and have gotten better. They are different from the suboccipital pains.

(*j*) The pulling or drawing-up feeling in the left eyebrow (not eyelid, apparently) is not accompanied by any visible drawing up, nor has any facial twitching been noticed; but she associates it with the wider palpebral fissure on that side which she has noticed. The feeling first occurred in 1917, and together with the trouble in the hand was the first thing noticed. It is somewhat better now.

(*k*) The suboccipital pains were first noticed the last of 1919. They have not troubled her much. They are caused by turning the head to the left or bending it back. After this the queer pain comes on, not intense nor sharp, more dull. It comes on with a rush as though blood or something rushed in. They are getting worse.

(*l*) The inequality of temperature sense in the teeth on the two sides of the mouth has been noticed for less than a year. This is getting a little worse. It consists of a hypersensitiveness of the left teeth to hot and cold. They are not over-sensitive to biting hard.

(*m*) The peculiar feeling in the left thigh appears to consist of a weak feeling similar to that in the left hand. After walking she has a lame feeling there, never severe. The weak feeling was first noticed in the last of 1919 and is getting worse.

(*n*) The buzzing in the left ear after stooping is very slight and does "not amount to anything."

(*o*) After stooping there is a feeling of rush of blood and things get dark. This was first noticed in July, 1919, and has not been noticed since last summer.

(*p*) Looking to the right feels uncomfortable in or around the left eye.

Birth.—The patient has been told by her mother that instruments were used at her birth and that a scar on her left temple is due to that.

Development.—She also has been told that she began to walk at sixteen months and to talk at about the usual time. She was fed Mellin's Food when a baby and suffered from malnutrition.

Injuries, Diseases, etc.—There is no history of injuries of importance to head or to other parts excepting possibly an injury to the left ankle three years ago in which it was wrenched badly and had to be bound up. Adenoids were removed at four years of age. There is no history of convulsions, loss of consciousness, paralysis or disturbances of speech. There is no history of headache. The patient only remembers having had three in her life including one severe one two years ago. She has had attacks of vomiting with nausea. The vomiting was forcible and preceded by nausea. This occurred every morning on getting up, before breakfast. There was no food in the vomitus, which was bright yellow and bitter. Sometimes she felt nausea again at about eleven in the morning, but did not vomit, after which she had no more nausea nor vomiting for the rest of the day. The vomiting began in October, 1920, and lasted till Christmas or January. It was attributed by the patient to some bad news she had received. For the last few years her stomach has been more easily upset by fatigue and there have been nausea and occasional quick vomiting but not frequent attacks like the above. This is attributed by the patient to the work she took up then on a farm. A severe bilious attack with headache in May, 1919, is recalled by the patient, the first vomiting with bright yellow bile.

In infancy, the patient had an otitis in one ear. Thinks it was the left ear. She had measles at two years, chickenpox and mumps at nine years. She never had diphtheria, influenza, malaria, meningitis, kidney trouble, whooping cough, pneumonia, rheumatism; scarlet fever, syphilis or typhoid.

Habits and General Physiological Status.—There is either no excessive or no use at all of tea, coffee, alcohol and tobacco. She sleeps well except that her sleep is interrupted by nocturia once a night, after which she does not fall asleep for an hour. The nocturia began last November. She has no appetite, the loss apparently beginning since 1917. She is never thirsty. Her bowels are regular. In 1919 she suffered from constipation. There is no history of fever nor of any significant change of weight. Menstruation has been very irregular, having occurred as follows: May, 1919, July 4, 1919, last of September, 1919, January, 1920. It was then regular till June 17, 1920, and there has been none since. When it occurs it lasts four to six days, the amount being small. She was never absolutely regular. There has been no dysmenorrhea. She had a leucorrhea in 1917. Dr. Marion Kenworthy, to whom we are indebted for the case, has inquired into this and thinks it was simply a catarrhal condition aggravated by standing a great deal.

Family History.—The patient knows of no miscarriages nor stillbirths by her mother, who has not lost any children after birth. She has one sister eleven years old, a bright healthy child, who had one convulsion (attack of stiffening) at two and one half years of age, attributed to an indiscretion of diet. Her father was drowned at forty-five years of age. Her mother is living and was well till last year. She worries and has "neuritis" and a "bad heart." She is forty-three years old. No similar trouble is known in the family. An aunt has some nervous trouble.

NEUROLOGICAL EXAMINATION

VOLUNTARY MOTOR SYSTEM. *Abnormal Attitudes and Deformities.*—No deformities were noted. When the upper extremities are extended in front, the left tends to assume a position lower and more outward than the right.

Gait.—The gait with the eyes open is somewhat irregular and ataxic. There is no change when walking on heels or toes but she walks more poorly on a straight line. She walks backwards rather better than forwards and runs better than walks. The Fournier tests showed disturbance but not as marked as would be expected. The greatest change was when walking with the eyes closed. She then staggered very badly.

Coordination.—The patient is right-handed. Standing with the feet together and eyes open, she shows some unsteadiness. On closing the eyes the unsteadiness is increased and she tends to fall to the left. This tendency is not altered by turning the head to the left or right but appears to be diminished by bending the head forward or backward. She can stand on the right foot but not on the left. The finger-to-nose test is performed normally with the right hand but the left shows ataxia and dysmetria, i.e., the nose is missed and the movement is performed with irregularities both lateral and vertical (non-motofacient) and in the plane of movement (motofacient and dysmetric). The performance is not very markedly affected by closing the eyes, but it is interesting to note that there is at times more improvement when the patient does it slowly with the eyes open and looking at the finger than when she does it rapidly looking at it, as compared with the eyes closed. In the former case there is apparently more time for correction by the eye and probably also by muscle-tendon sensations.

The fingers-to-thumb test is performed normally with the right, ataxically with the left fingers.

Pointing and pass-pointing is performed normally with the right upper extremity. With the left there is a marked tendency to deviate to the left. Quite often the movement begins with a marked deviation to the left and is then brought back to the proper point as though, aware by muscle-tendon sense of the deviation, the patient corrects it.

The heel-to-opposite-knee test is performed normally on the right. On the left the foot overreaches, often misses the knee and passes along the shin with a very wabbly movement.

Adiadochokinesis is present on the left. Besides the above tests dysmetria is also shown by stopping abruptly in elevating and bringing the left arm down in front, in supination, pronation, etc.

If the arms are extended and the palms of the hands rest on those of the examiner, the patient being told to let the weight rest on the examiner's hands, sudden removal of the examiner's hands causes the left hand of the patient to fall further than the right. When the patient's arms and hands are extended forward, eyes closed, a smart tap elicits more rebound on the left. Sudden release of the flexed forearm elicits more rebound on the left.

When the arms are extended in front, eyes closed, the left arm tends to sink and occasionally deviate outward.

Skilled Acts.—The handwriting shows no abnormality. It is rather irregular but was performed quickly and easily. The speech in conversation and test phrases is normal.

Abnormal Involuntary Movements.—When the upper extremities are extended in front there are irregular jerky movements of the left fingers. These appear to be present, in much less degree, in the right hand.

Reflexes.—Deep. These are active on both sides. The left patellar has perhaps somewhat more amplitude than the right and is pendulous. There are no crossed or uncrossed tibio-adductor (periosteal) reflexes and no ankle clonus.

Superficial. The abdominals are fairly active and equal. There is no Babinski nor any of its modifications.

Muscle Strength.—In general on the left side there appears to be slightly less strength than on the right, hardly more perhaps than the normal difference. Some of the opposed passive movements, however, show remissions of tension followed by quick corrective contractions, i.e., the position of the part is not held steadily.

Muscle Status.—The left upper extremity shows a slight hypotonia as compared with the right—more flabby, but not markedly more extensible except the thumb, which the patient says has always been "double-jointed."

Abnormal Associated Movements.—There is no sign of Hoffman, no thumb sign of Klippel-Weil, no tibialis sign of Strümpell, and no inability, when recumbent, to hold up both lower extremities (Grasset-Bychowski). On sitting up, or trying to, the left lower extremity is elevated more than the right.

GENERAL SENSORY. *Face.*—In general cotton tickles more on the right, but in the left maxillary region there is a peculiar numb, tingling sensation. The left half of the tongue shows marked diminution of sensibility to contact (toothpick), to pin point, and to cool and warm. The floor of the mouth shows diminution to contact. Pain and temperature not tested. The floor of the mouth outside the gums, the inner surface of the lower lip, the oral surface of the cheeks and the outer surface of the chin appear to show only a slight diminution on the left. The roof of the mouth appears to show no change.

There was not a thorough sensory examination made of the trunk, but the extremities showed no changes in touch nor of the sense of position and movement. There is possibly a very slight diminution of temperature sense (warm) on the right (hand and neck), but it is too slight and doubtful to constitute a reliable diagnostic sign.

CRANIAL NERVES. *Olfactory.*—Smell is normal (oil of cloves).

Optic and oculomotor apparatus.—The following is a report by Dr. Conrad Berens of an examination made by him:

The patient complained that her eyes jerked and things became blurred when she looked to the left. There was double vision at times. Her previous eye history was negative except that she had worn glasses for one year at the age of sixteen. Her present eye trouble commenced in 1919 with blurring of objects upon looking to the left. She has noticed vertical diplopia at times during the last year.

Vision in the right eye, 20/20, corrected under homatropine to 20/20 plus by a +.25 cylinder axis 75°. Vision in the left eye, 20/30 plus corrected by a +.25 cylinder axis 90° to 20/20 plus.

The pupillary distance was 64 millimeters; near point of convergence, 160 millimeters from the cornea (it should be under 75 millimeters to be normal). The near point of accommodation from the cornea, in the right eye is 120 millimeters, and in the left eye 128 millimeters (this is nearly normal for her age, 110 millimeters being absolutely normal).

The muscle balance showed for 6 meters 1 prism diopter of exophoria and 6 prism diopters of right hyperphoria. Nystagmus was the most marked sign revealed by inspection. With the eyes fixed for distance, the right eye showed rotary nystagmus, irregularly rhythmical with a slight occasional vertical jump, slow movement clockwise. The left eye, rotary nystagmus with an occasional vertical jump, slightly greater amplitude than the right eye; slow movement clockwise. Rate, ninety a minute for rotary component and ten a minute for the irregular vertical component. Upon looking down the nystagmus disappears. Looking up and to the right, horizontal amplitude 3 millimeters, 140 a minute; slow component to the left. Looking up, slight rotary component noted. Eyes up and to the left, horizontal, vertical and slight rotary nystagmus; small amplitude, rate 130 a minute. To the left, horizontal nystagmus, 130 a minute; slow component to right, amplitude, 6 millimeters; slight rotary component.

Tension normal in both eyes.

Fields for form and color. Right eye normal. The field for form and color in the left eye was slightly contracted above and temporally (possibly due to the nystagmus). The blind spots were normal at 75 centimeters.

The eyelids and lachrymal apparatus were normal except that the left palpebral fissure was larger than the right and there was a slight Stellwag sign on the left side. She winked more on the right.

The ocular movements showed spasms and weakness of several muscles, but the only constant insufficiency was that of the left external rectus.

Diplopia fields plotted on the tangent screen showed vertical diplopia in all fields, more marked looking to the left but not increased by looking up or down. The right was the higher eye. The conjunctiva was normal. Left cornea slightly insensitive to tactile stimulation.

Irides reacted normally to direct and indirect light, accommodation and convergence, and to ciliospinal reflex stimulation.

Pupils fixed for 6 m. in a moderate illumination, 4 mm. in diameter, equal and regular in outline.

Media and fundi were normal.

Acoustic Nerve.—Hearing is diminished on the left, both air and bone conduction being involved.

Dr. Randal Hoyt was asked to test the acoustic, especially the vestibular portion. The following is his report. It must be noted that his suggestions as to localization were based on his vestibular findings considered by themselves:

Auditory Status.—Tinnitus was not present. Whisper, right 20/20, left 10/20. Tuning fork tests, C. 128, right 20/20, left 10/20 in both air and bone conduction. A.C. greater than B.C. C.512 not heard in left ear.

Vestibular Status (pre-rotatory).—Spontaneous vertigo was not present. Nystagmus, fine, on looking to right; coarse, on looking to left. Balance: sinistropulsion (?); retropulsion on walking backwards. Pointing tests, dysmetria with left; no deviatory movements.

Rotation Tests.—Series 1. Upright Position: (1) Direction of rotation is recognized instantly. (2) Duration of movement after sensation of having stopped has been signified, right greater than left. (3) Sensation of rotating in reverse direction is induced on right by two revolutions, on left by one-half revolution.

Series 2. Lateral Falling Position (head bent forwards, face parallel with floor): (1) Falling in direction of rotation is induced by sixteen revolutions to right and three to left. (2) Sensation of falling opposite to direction of rotation is not induced by revolution to right or left.

Series 3. Dorsoventrad Falling Position (head bent forwards, face turned to left): (1) Falling forwards is induced by three revolutions to right and also three to left. (2) Sensation of falling backwards is induced by three revolutions to the left but not to the right. (3) Sensation of falling forwards is induced by three revolutions to the left but not to the right. Slight nausea induced by falling tests. No vomiting induced by tests. No pallor induced by tests.

Nystagmus.— Rotation to right causes coarse nystagmus, already present on looking to left, to take character of vestibular (fine) nystagmus. It completely inhibits the fine nystagmus that is present when patient looks to right. Rotation to left causes fine nystagmus, already present on looking to right, to become more intense and then coarse for a few seconds. It does not influence the coarse nystagmus that is present when patient looks to left.

Analysis.—The hearing tests show that the left auditory nerve is involved; they also show that it has not been completely destroyed. The fine nystagmus on looking to the right is similar in character to that produced by rotation to the left in normal individuals. It is what might be expected were impulses from the right labyrinth predominant over those coming from the left. The inhibition of this nystagmus by rotation to the right indicates that this discrepancy has been equalized. The fact that vertigo to the right can be induced by one-half revolution to the left, is accounted for on the same basis.

The failure to induce pass-pointing by rotation to the right indicates a block in the left vestibular nerve. The delay in induced dextropulsion, and the absence of induced sinistropulsive vertigo, are in keeping with the above. The absence of dextropulsive vertigo, after rotation to the left, however, is a dissociation that cannot be explained on the basis of a vestibular nerve lesion.

With the head bent forward and face turned to the left, the right posterior and the left superior canals are in the plane of rotation. Rotation to the left stimulates the right posterior canal more than the left superior. The resultant direction of movement while normal was not of the usual intensity. The presence of sinistropulsive instead of retropulsive vertigo, is due probably to the fact that impulses coming from the left superior canal were delayed.

Rotation to the right (in this position) stimulates the left superior canal more than the right posterior, and the failure to induce either movement or sensation are in keeping with a lesion in the left vestibular nerve.

With the head bent forward and the face turned to the left the left posterior and the right superior canals are in the plane of rotation. Rotation to the right stimulates the left posterior canal more than the right superior; and the absence of motor or sensory responses to such rotation are in keeping with a lesion in the left vestibular nerve.

Rotation to the left (in this position) stimulates the right superior canal more than the left posterior. The fact that sinistropulsion instead of retropulsion should have followed such rotation may be explained on the basis that impulses from the left posterior canal were delayed. That dextropulsive vertigo should not have accompanied falling to the left, cannot be explained by a vestibular nerve lesion.

Thus, whereas the majority of the above tests indicate a lesion of the left vestibular nerve, the spontaneous coarse nystagmus on looking to the left, coupled with the fact that it is not inhibited by rotation to the left (i.e., by stimulation of the right horizontal canal) and the fact that there are evidences of dissociation between the motor and sensory effects of rotation, indicate that there is involvement of parts other than the vestibular nerve.

Dissociation of the sensorimotor effects of rotation suggests involvement of the cerebral cortex. In this case, the only part of the cortex that is in relation with the peripheral nerves so obviously involved, is the left temporal lobe.

On this account, and because none of the involved nerves is completely destroyed, my own impression is that there is a lesion, which primarily involves the left temporal lobe, but which by extension downwards has secondarily involved some of the cranial nerves.

Trigeminus nerve.—The corneal reflex is present on the right and diminished on the left.

The jaw deviates to the left on opening the mouth. The patient can bite harder on the right than on the left.

Facial Nerve.—Bitter, sweet and sour were tasted on the right but not on the left. Some allowance must be made for possible confusion on account of other lingual sensory changes.

There is a left peripheral facial paresis, not very marked.

Glossopharyngeus and Vagus Nerves.—The patient thought she did not taste the bitter substance on the left side at all when the tongue was retracted, apparently indicating that taste may be also disturbed on the back of the tongue, left side. This, however, cannot be regarded as certain. The vocal cords were examined by Dr. Osgood and found to be normal.

Hypoglossus.—The tongue is protruded without deviation though it is nearer the left angle of the mouth.

MENTAL STATUS.—The impression during examination is that the patient is normal as to intelligence, emotion, attention and memory. It does not appear that the feelings of dullness and stupidity of which she complains can have affected her noticeably.

CRANIAL MORPHOLOGY.—Percussion on the left side elicits a different sensation from the right.

SYSTEMIC. *Tegumentary.*—The patient

called attention to the fact that she perspired more, as shown by greater dampness of the garments, under the right than under the left axilla.

Cardiovascular.—The heart is practically normal. It is a little enlarged to the right. There is a slight mitral presystolic murmur and accentuation of the second pulmonic sound which has almost a pulmonic ring.

Laboratory Tests.—The blood Wassermann test is negative. The spinal fluid has not been tested, it being thought wiser to defer this.

SUMMARY

The physical signs thus clearly indicate a lesion, or lesions, affecting the left fifth, seventh and eighth cranial nerves, the left cerebellum and possibly the left sixth nerve.

It will be agreed, probably, that the objective signs point to a lesion in the left cerebello-pontine angle or its vicinity. This is, perhaps, even more true of the subjective signs—the complaints—which read remarkably like the various cases reported by Cushing. While they are quite typical of a cerebello-pontine angle neoplasm there are, nevertheless, some other possibilities to be considered.

In the first place there is to be noted the predominance of cerebellar symptoms, reenforced probably by vestibular disturbances. This is quite typical; but it must be remembered that this is a rather early case, if a neoplasm at least without any marked intracranial pressure, and if extra-cerebellar perhaps the cerebellar symptoms would not be expected to be quite so conspicuous. The possibility of an intra-cerebellar lesion should thus be considered. There is no certain differentiation, but it would hardly seem that, in this case, the symptoms, especially those due to the cranial nerves, would be quite so definite and unilateral, and it would seem that the general symptoms, especially headache and vomiting, would be more severe were the latter the case.

The great increase in unsteadiness of gait when the eyes are closed is interesting and not characteristically cerebellar. Might this be due to the middle peduncle being involved?

The nystagmus is, owing to its complicated character, difficult to interpret precisely. It follows the rules of being coarser with the eyes directed to the side of the lesion and may, perhaps, be best regarded as a mixture of vestibular and cerebellar elements.

If we regard the lesion as a cerebello-pontine angle neoplasm, the further question arises whether it is an acoustic nerve neoplasm or some other, e.g., a lateral recess, growth. In favor of the latter is perhaps the fact that hearing has not ben so severely impaired, nor possibly noticed so long, as would be expected with an acoustic nerve growth. The absence of marked tinnitus would point in the same direction.

An interesting symptom is the greater involvement of the third division of the fifth nerve. This would apparently indicate a pressure on the nerve from its caudal aspect as it leaves the cranium. This and the absence of vocal cord symptoms may indicate the position of the focal lesion or neoplasm to be somewhat in front of the cerebello-pontine angle and on the middle cerebellar peduncle.

Multiple sclerosis may be mentioned, but excluded on account of lack of signs of multiplicity of lesions and absence of pyramidal signs.

Epidemic encephalitis may be considered, but the consistent focal symptoms and steady progress of the disease as well as the fact that the group does not resemble the encephalitic groups may be taken to exclude this possibility.

DISCUSSION

Dr. Stookey thought that the case was very interesting because of the relatively few 8th nerve signs, and also on account of the chronological appearance of the signs. All of the 5th nerve symptoms seem to have come on before the 7th and 8th, and seem to predominate. He thought a tumor of the Gasserian ganglion had to be considered. Such a tumor usually causes more severe pain, but he did not believe that it should be excluded.

Also very interesting were the marked cerebellar symptoms. It seems rather difficult to combine the 5th nerve and the cerebellar symptoms. It is not a typical tumor of the 8th nerve, because the 8th nerve signs are very slight. A tumor of the 5th nerve has to be considered. He wondered whether examination had been made of other parts of the body. Frequently associated with this sort of condition there are other tumors in the body. He had seen one such case in which two tumors were found in the upper extremities.

DR. EVANS stated that a number of stereoscopic *x*-ray plates had been taken of the posterior fossa, and nothing could be seen on them suggestive of tumor.

DR. OSNATO thought that the most remarkable thing about the case was the absence of the ordinary constitutional signs, if one may call them such, of brain tumor—headache, vomiting, etc. There is no condition in which such severe headaches occur as in brain tumors, especially cerebello-pontine tumors. There are also no mental changes. If this is a tumor, it seemed to him that it must be a slow gliomatous one, infiltrating, and that the brain must be slowly accommodating itself to its growth.

DR. CASAMAJOR felt that the case was puzzling, there being no headache and no choked disc. He has found that the only posterior fossa tumors that do not give these signs are those of the pontine region. Here the 5th nerve was the one most affected, particularly the motor part of the 5th, secondly the cerebellum, and, much less, the 7th nerve; the 8th nerve was hardly affected at all. These facts would make one think that this lesion began somewhat further forward than the cerebellopontine angle. He wondered whether the cerebellar signs could be attributed to the pons.

He thought that the lesion was probably a tumor, as there was nothing else he knew of that went along with gradually increasing symptoms as this has done. He thought the lesion was very likely cephalad of the cerebellopontine angle, possibly lying along the side of the pons, and in its extension backward was now beginning to involve the cerebellum, so that later on, symptoms of a cerebello-pontine angle tumor might occur, but he doubted whether they could be called such at present.

DR. STRONG considered that the 8th nerve was certainly involved because there was a decided impairment in hearing on the left side.

He did not agree with Dr. Casamajor as to the tumor being in the pons region because he did not think there could be a lesion anywhere in the pons without bilateral symptoms. Besides a lesion in the pons is almost sure to give pyramidal symptoms and later some sensory symptoms. He thought that the evidence was rather against an acusticus tumor, but not against a cerebello-pontine tumor. The absence of choked disc was not necessarily against the theory of a cerebello-pontine tumor.

DR. TILNEY said that we should seriously consider what is the best course to follow in all such cases where diagnosis of brain tumor is somewhat conjectural. Recently he saw what seemed to be a tumor with cerebellar symptoms predominating. Dr. Elsberg examined the patient and admitted that there were signs of intracranial pressure. He also found the abdominal reflexes diminished and came to the conclusion that the case was one of multiple sclerosis, and did not advise operation on that ground. Later the patient went to Dr. Cushing who maintained that there was a cerebellar tumor and that the man should be operated at once. An operation was performed and all that was found was a thickening in the arachnoid.

Dr. Tilney felt that neurologists should be a little more courageous than they have been in advising operations. Surgeons were making many cranial exploratory operations without harm to the patients. If there is a tumor, of course the time to get it out is before it has produced too much damage. He considered that an exploratory operation is indicated when the symptoms are as pronounced as they are in the case under discussion. An abdominal surgeon in dealing with a case having symptoms of anything like such severity, would operate at once. The brain surgeon should be entitled to similar advantages of early interference since delay almost invariably led to permanent destructive changes in the brain. He advised in this case an immediate subtentorial exploration in the expectation of discovering a cerebello-pontine lesion.

Dr. Casamajor agreed with Dr. Tilney on the point of early operation and said that if we are going to wait until we get a choked disc, it is usually too late to operate. If we continue to wait before operation as long as we do now, he feared that neurological surgery was going to be as hopeless as it has been in the past. Of course one does not like to recommend craniotomy in a young individual, on account of the high mortality; but he wondered whether this high mortality was not due to the fact that the operations are usually done too late. Personally, he would rather see an exploratory done early in a case of this kind. Usually cerebello-pontine tumors do not occur as early as this, and it was just by a fortunate circumstance that this patient came to the clinic. Dr. Casamajor thought that as many opinions as possible should be obtained, not only neurological, but also surgical. He recommended that the patient should go to the hospital for several weeks for observation.

Later Comment: The subsequent operation apparently showed that the objections to a cerebello-pontine angle localization, such as the lack of especially severe involvement of the acoustic and absence of marked tinnitus, together with the pronounced cerebellar symptoms, did not take sufficient account of the possibility of a cerebello-pontine angle growth of such a yielding character as not to give excessive blocking and irritation of the acoustic nerve.

REPORT OF OPERATION BY DR. ELSBERG

March 5th, 1921:

Suboccipital craniotomy for lesion (cyst?) in left cerebello-pontine angle—by Dr. Elsberg.

Ether anesthesia. Typical suboccipital craniotomy through cross-bow incision. Hemorrhage moderate. Incision of dura; only moderate increase of intracranial pressure. Cisterna magna of moderate size and moderately distended. Aspiration left cerebellar lobe negative. Deep in the left cerebello-pontine angle covering the nerves in the angle was a cystic mass, its surface partly roughened, cloudy and partly translucent. The mass did not feel solid, and during manipulations it ruptured, allowing some clear fluid to escape. There was no evidence of a tumor underneath. Nothing further done. Closure of wound in the usual manner. Convalescence from operation uncomplicated. Sutures removed on seventh day; primary union. No increase of cerebellar symptoms due to operative manipulations.

DYSTONIA MUSCULORUM DEFORMANS

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NEW YORK CITY

One of the most interesting of the extrapyramidal motor disturbances is the syndrome which has been variously named *dystonia musculorum deformans, tonic torsion neurosis, progressive torsion spasm* and *tortipelvis*. This condition has been so well described by Oppenheim, Hunt, Ziehen, Bernstein and others, that a full description of the subject would be superfluous. Suffice it to say that it is a motor disturbance which is characterized by alterations in tone of the musculature of the extremities and trunk, from extreme flaccidity to marked hypertonicity plus more or less systematized torsion—twisting movements of the parts involved. Its onset is during adolescence, and usually at first it is limited to one of the lower extremities, gradually progressing until the trunk and all extremities are equally involved. As a result of these spasms there are deformities of the skeleton, as curvature of the spine, with tilting of the pelvis and distortion of the chest, deformities of the feet and occasionally fixation of a joint through muscular contractures. The location of the lesion is probably in the corpus striatum, but this is at present largely speculative, as few cases have come to autopsy. Thomalla found degenerative changes in the globus pallidus in one of his cases.

HISTORY

Patient: Miss E. S., fourteen years of age.

Chief Complaint.—Progressive twitchings and involuntary movements with weakness; duration five and a half years.

Family History.—There is no history of syphilis or other chronic disease. The mother had tuberculosis of the ankle with subsequent amputation below the knee, followed by a tuberculous cervical gland which was also removed when the patient was eleven months of age. She has been entirely well since and never at any time has she had trouble with her lungs. The father is living and well. One sister is rather delicate, but well and of average mentality. The father is Scotch; the mother is Austrian and of the Jewish faith.

Previous History.—Birth was normal. She was always a bright and healthy child. When at school for the two years prior to the onset of the present illness, she progressed well and skipped one class. She had a mild attack of whooping cough at five years; measles without complications at six; scarlet fever at eight. Shortly before the attack of scarlet fever she had pains in her legs which lasted a few days but did not confine her to bed. The condition was diagnosed by her physician as "growing pains."

Ears, eyes, nose, throat, cardiac, respiratory and gastro-intestinal history negative. She has never menstruated. The neuromuscular history was negative prior to the onset of the present illness.

Present Illness.—One morning, without any premonitory symptoms, having passed a quiet night, it was noted by her mother that she shrugged her shoulders spasmodically. This was five months after the attack of scarlet fever and she was at that time eight years of age. Her physician considered the condition to be chorea and kept her in bed in a dark room for a week. This treatment afforded no relief, and she was allowed to return to school for two months. At this time her work was so excellent that she was allowed to skip a class. A little later her left foot started to turn in with stiffening of the muscles of the toes and foot. The muscles of the back became weak so that the trunk was bent slightly forward. She was taken out of school and sent to the Neurological Institute, New York City, where she stayed one month. They removed five of her first set of teeth, tonsils and adenoids, without any relief of the symptoms, and no diagnosis was made.

Gradually she stopped shrugging her shoulders and it was noted that her foot had short periods of being relaxed. At that time her eyes started "rolling around" and she commenced to lose control of her legs and to have difficulty in walking. This was nine months after the onset. She then went to the Red Cross Hospital for eleven weeks, where blood and spinal fluid examinations and *x*-ray pictures of the spine were made, all of which were said to be negative, and no diagnosis was offered. At about this time her voice began to quaver and in six months the change was so marked that strangers could not understand her. This condition persisted until one year ago, when her voice became clearer again and has remained so. After this the legs grew weak and uncontrollable, she also lost control of the right arm, which took on choreic movements. Later her neck and left arm became involved and her back muscles showed unbalance resulting in curvature of the spine. For about a year (three and a half years ago) she had no control of her arms at all. For the last two and a half years, however, she has had better control of her arms and head. Her mind has apparently not been noticeably affected. Her memory for the past and present is good and she seems bright and intelligent. She has never had incontinence, but has been obliged to use laxatives for the last three and a half years, during which time she has been confined to bed. She has had no convulsions, no vomiting, no fever and no pain.

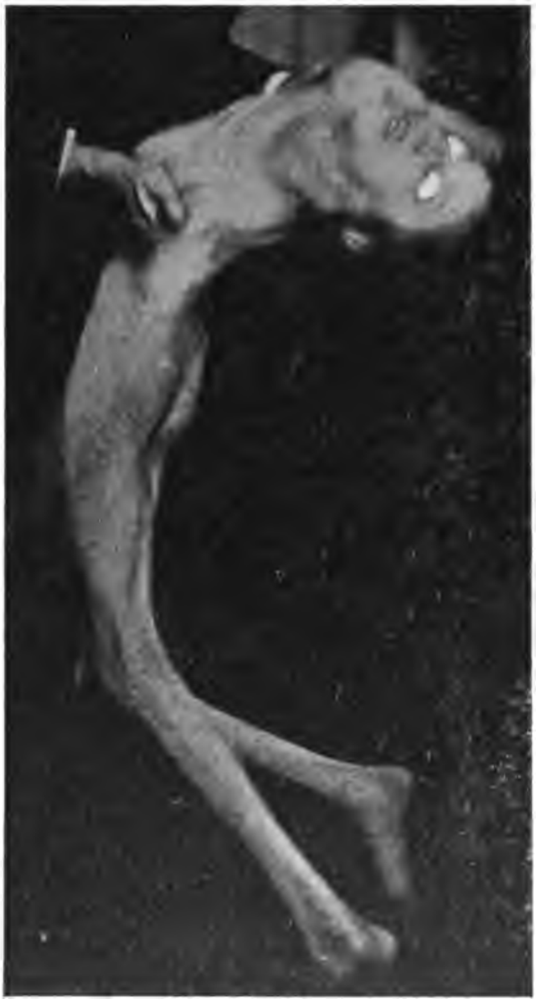


FIGURE 1.

PHYSICAL EXAMINATION

General Appearance.—The patient is lying grotesquely in bed, with her head and extremities in constant motion. The head is flexed backward with marked hypertrophy of the neck muscles, and every few seconds there is either a rotary, forward or backward jerk. The body is quite deformed and misshapen. There is a marked right thoracic scoliosis and some kyphosis, as if the vertebrae were also rotated to the right. The chest is badly deformed, being flat on the left side and having a sharp edge in the left axillary line.

Head.—The head, well formed, is quite retracted. While she is awake it is in constant motion.

The pupils are equal and normal. The conjunctivae are slightly pale, smooth and not injected. There is no nystagmus. The ocular movements are under perfect control of the will and the eyes are kept quite steady. Vision is normal.

The ear drums are negative. The nose is normal and there is no discharge. The mouth is quite clean, teeth in fair condition. The tonsils are absent except for several small tags of tissue in the left fossa.

Speech is jerky and articulation is labored and at times indistinct. The speech disturbance seems to be due to the jerking of the muscles of the neck.

The neck is held stiffly backwards and there is considerable hypertrophy of the neck muscles. The superficial veins of the neck are engorged. No glands are made out and the thyroid is not enlarged.

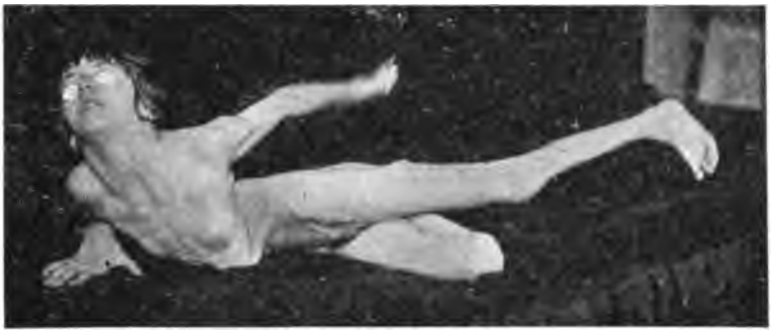


FIGURE 2.

The thorax is exceedingly misshapen. The left thoracic wall is retracted at a sharp axillary angle of the ribs, while there is bulging of the chest to the right, as would be expected with such a marked degree of scoliosis. Expansion: There is a fair amplitude but it is also more marked on the right side. Percussion note is resonant throughout, but the angulations of the chest make the changes in pitch unreliable. The breath sounds are vesicular. There are no râles.

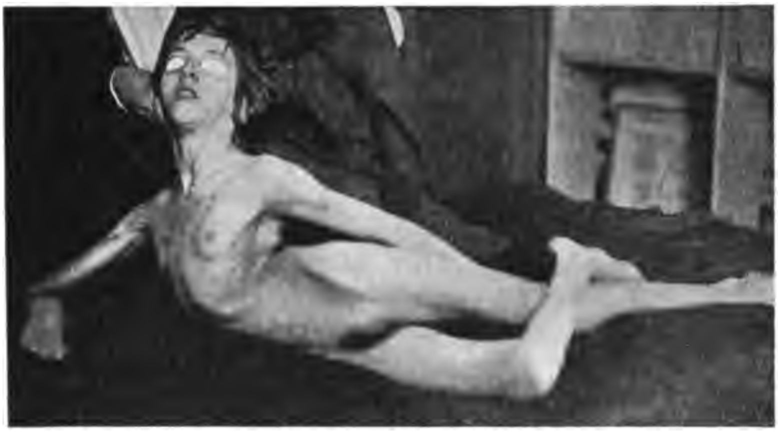


FIGURE 3.

Heart.—It is impossible to localize the border by percussion, but there is no evidence of cardiac hypertrophy. The apex beat is not visible. The rate is 90 and perfectly regular. There are no murmurs.

The abdomen is soft, retracted and thin-walled. Nothing remarkable is found on palpation. Tenderness, masses and signs of fluid are absent. The liver and spleen are not felt.

Genitalia are normal to external examination.

NEUROLOGICAL EXAMINATION

November 9, 1921

Abnormal Attitudes and Deformities.—The patient lies on the bed in the most bizarre and contorted positions. There is constant movement of the muscles of the extremities and trunk so that no position is maintained for more than a few minutes. The legs are frequently flexed on the thighs so that the heels are in apposition with the buttock while the thighs are extended. The pelvis is tilted to the left and the spine rotated so that the ilium is nearly in the axilla. The spine is markedly scoliotic and frequently arched backward with the head and neck bent to produce extreme opisthotonos. Though the movement is continuous and the long axis of the body is as frequently in the transverse position on the bed as it is in the longitudinal, she never falls off the bed. The deformities present are an inversion of the left foot (but without tendency to varus or valgus), marked spinal scoliosis, fixation of the left hip, and the inability to extend fully either leg at the knee. She is unable to stand or to maintain the sitting posture.

Gait cannot be tested on account of the inability to stand even if supported.

Coordination.—On account of the constant movements it is impossible to determine the condition of coordination, dysmetria or adiadochokinesis.

Skilled Acts.—There is no dyspraxia. Dysarthria is marked; at times it is extremely difficult to understand what she is trying to say. This seems to be a dystonic phenomenon due to the abnormal conditions in the muscles of the larynx, palate and lips. Handwriting: It is impossible for her to write.

Reflexes, Deep.—The deep reflexes seem to be present but are extremely difficult to elicit because of the constant muscle tension. There is no difference in reaction between the two sides.

Reflexes, Superficial.—The abdominal reflexes are present and the plantar reflex is of the normal flexor type.

Abnormal Involuntary Movements are not convulsive, tremorous or choreiform, but are tonic, twisting, torsion movements of large muscle groups involving mainly the trunk and proximal portions of the extremities. They are, however, present in the hands and feet but not in the muscles of the face or eyes. The movements are somewhat similar to grand athetoses though no definite formulae are constantly followed.

Muscle Strength.—The muscle strength is good, the muscular power being under the control of the will.

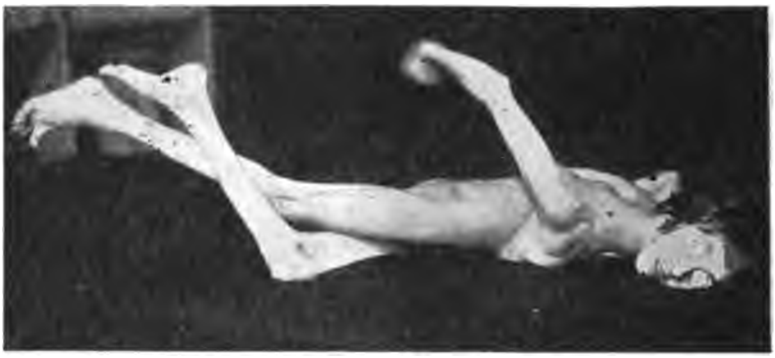


FIGURE 4.

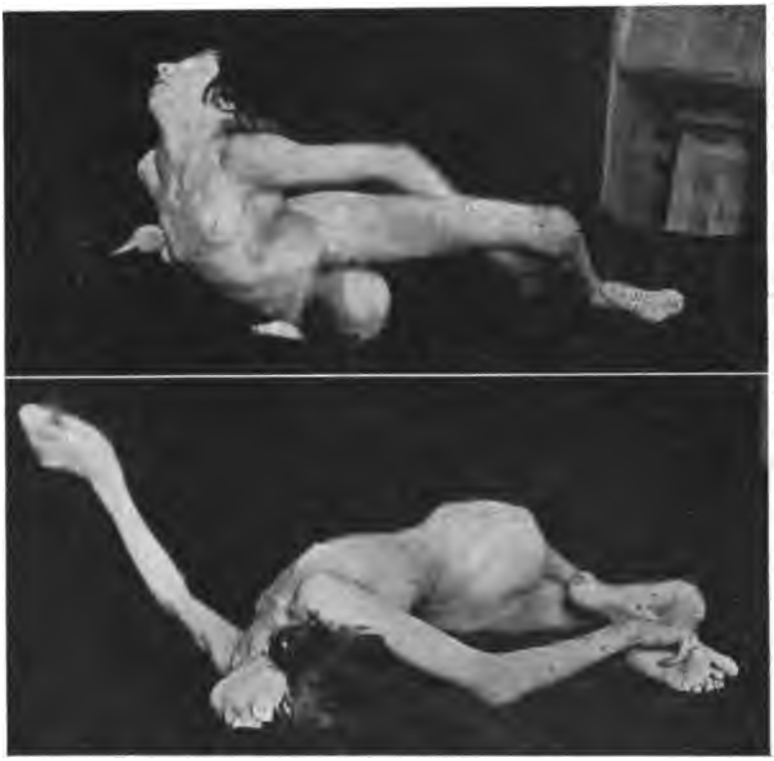
Muscle Status.—The muscles of the extremities are all small, firm, and for the most part rigid, due to the tonic contractions. The muscle tone varies markedly, however, from an extreme tension to complete flaccid relaxation. Irritability is normal. Myoidema is absent. The paradoxical phenomenon of dystonia (Hunt) is present in both wrists and ankles. To elicit this sign the extremity is flexed and the patient directed to extend it. On attempting to do so further flexion is produced first, and it is only after a perceptible interval of time that extension occurs.

Nerve Status.—Normal.

General Sensory.—It is astonishing that there is no subjective pain, considering the extreme deforming positions so constantly assumed. Pain is present, however, if the movements are passively restricted. Touch, including localization and discrimination, is normal. Pain, temperature, vibratory, pressure, muscle tendon sense and stereognostic perception are intact.

CRANIAL NERVES AND HEAD. *Olfactory Nerve and Nose.*—Smell is normal in each nostril.

Optic and Oculomotor Apparatus.—Visual acuity is good. The pupils are markedly dilated; they react actively to light and on accommodation. There is no exophthalmos, enophthalmos or ptosis. Strabismus and nystagmus are absent. The conjugated eye movements are normal.



FIGURES 5 AND 6.

Acoustic Nerve and Ear.—A watch is heard three feet from either ear. Air conduction is greater than bone on each side.

Trigeminus Nerve and Mouth. — The movements of the jaw in all directions are equal. The corneal reflexes are present.

Facial Nerve and Face.—Taste was not tested. There is no paralysis of either the upper or lower facial muscle groups.

Glossopharyngeus and Vagus Nerves, Pharynx and Larynx.—The uvula is in the midline. The palatal and pharyngeal reflexes are present. Swallowing at times is said to be difficult but this is believed to be due to the sharp retraction of the neck and movements of the head.

Spinal Accessory Nerve.—Normal.

Hypoglossus Nerve and Tongue.—There is no evidence of paralysis.

Summary of Neurological Examination.—The neurological examination is negative except for the tonic torsion movements and alternating hyper- and hypotonic muscular disturbance.

LABORATORY EXAMINATIONS

Urine examination: November 7, 1921.

Color	Amber
Specific Gravity	1.035
Reaction	Acid
Precipitate	Slight flocculent
Albumin	none
Glucose	none
Microscopical examination	negative

Blood Examination: November 10, 1921.

Hemoglobin	75%
White blood cells	9,400
Polymorphonuclear leucocytes	41%
Small lymphocytes	55%
Transitionals	2%
Eosinophiles	1%
Basophiles	1%

A smear shows the platelets tremendously increased.

The red cells are of normal morphology.

CONCLUSION

This case is of interest for several reasons: Nearly all of the recorded patients have been Jews; this patient's mother only was of Jewish blood. In most instances the first disturbance has been noted in one of the lower extremities, while this patient's symptoms commenced in the trunk with a shrugging of the shoulders. In but few of the reported cases has there been noted disturbance in the musculature innervated by the cranial nerves. In those of Bernstein and Keschner there were disturbances of speech which were supposed to be due to a dystonic condition of the muscles concerned in articulation. In Hunt's discussion of Bernstein's case he believes that the dysarthria was secondary to involvement of the neck muscles. In Keschner's case there were involuntary rolling movements of the tongue which became more marked on attempts to speak. In the patient whose history is here recorded, nine months after the onset the voice began to quaver, and six months later the disturbance was so marked that her speech was almost unintelligible. This condition persisted for over three years and then improved. Nine months after the onset there were said to have been involuntary ocular movements. These, however, persisted only a short time and were not present while the patient was under my observation.

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ATYPICAL FORMS OF EPIDEMIC ENCEPHALITIS

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The recent epidemic of encephalitis has given neurologists an unparallelled opportunity to study the various morbid phenomena resulting from the effects of the disease. Every type of neurologic abnormality has been encountered—syndromes of well-known orthodox types of nervous disorders and innumerable sub-types difficult of exact classification. As a result of the encephalitic disease—both in the manifestations of the acute phase and its sequelae—a revision of our conception of the causes of many neurologic disorders will have to be made. This has been pointed out by numerous authors who have studied the affection in all its phases. Many hitherto curious and obscure nervous affections can thus readily lend themselves to interpretation in the light of the present epidemic. There is no doubt that the disease has always existed in sporadic and endemic forms but has escaped recognition. The past epidemic with its bizarre and varied symptomatology has indeed left a rich heritage to future neurologic thought. The knowledge thus gained will enrich organic neurology and profoundly influence current notions and conceptions of the causes and mechanism of abnormal neurologic behavior.

While the major types of a disease process are easy of recognition during an epidemic, atypical or abortive forms quite often escape recognition, unless special study is made of them and all diagnostic aids used in an endeavor to give these cases a proper classification. This is particularly true of epidemic encephalitis. The gross clinical syndromes indicative of the affection needed little diagnostic acumen for their recognition, while the atypical forms, such as will be enumerated in the body of this paper, required considerable clinical and laboratory study for a proper interpretation. In retrospect, we are frank to confess (as indeed most neurologists are) that a not inconsiderable number of these atypical cases came under observation and were not recognized as variants of epidemic encephalitis. The paucity of symptoms, the evident insignificance of the complaints on the part of the patients, and the failure on the part of the observer to evaluate properly these minor manifestations, were all factors in permitting the diagnosis to slip through one's fingers, so to speak.

In a search of the literature (which has certainly done full justice to the epidemic in describing every possible neurologic and psychiatric syndrome) very little effort has evidently been made to describe atypical forms of the affection. All writers have given us excellent descriptions of the gross manifestations—the lethargy, fever, cranial nerve palsies, syndromes of basal ganglia involvement, etc., without further attention to abortive or atypical forms. The French, however, are a notable exception. With their marked clinical acumen they have described numerous variants of the affection in which all the phenomena that we are wont to associate with the disease were absent. These cases, they argued, could not be based on the assumption of a previous luetic infection, although the symptomatology very often pointed in that direction. Study of the spinal fluid enabled them to differentiate these cases and definitely to establish their identity in relation to epidemic encephalitis. We have been able through a study of six cases coming under observation to corroborate these findings. The number, while admittedly small, shows a varied symptomatology in which the diagnosis was made possible only by a study of the spinal fluids.

Because of our desire to call particular attention to the atypical symptoms which presented themselves in our cases, we have intentionally omitted unimportant details and have laid stress on positive findings only.

CASE I. G. R., age fifty-eight, male, no occupation (from the service of Dr. H. W. Frauenthal at the Hospital for Joint Diseases), whose previous history was unimportant, was stricken rather suddenly while attempting to mount a flight of steps. His legs suddenly gave way and he was precipitated to the ground, bruising his elbow considerably. He attempted to get up and apparently was momentarily successful, only to fall again. He was brought home and thence to the hospital, where the examination revealed an incomplete paraplegia. There was also urinary retention, and catheterization had to be resorted to at intervals to empty his bladder. He as able to stand up for a minute or so but was unable to walk any distance. The examination revealed a partial monoplegia of the right leg with a sustained ankle clonus and Babinski phenomenon. The deep reflexes of the lower extremities were equal and hyperactive. There was marked muscle sensitiveness in the affected extremity without demonstrable objective sensory changes. A transitory clonus was present in the other extremity. The patient appeared otherwise in good health and showed no disturbances in pulse or temeprature. Lumbar puncture revealed a fluid as follows:

Pressure	moderately increased
Cells	18 per cm.
Globulin	two plus
Sugar	reduction increased
Wassermann	negative in 2.0 c.c.
Colloidal gold	0111200000.

There has been no improvement in his condition to date.

CASE II. N. S., an attorney, forty years of age, whose previous history was absolutely negative, as the result of a severe financial reverse suffered a marked psychic episode and was unable to adjust himself to this unfortunate turn in his otherwise tranquil affairs. He slept poorly, became depressed and lost his appetite. This continued for three weeks, when he was awakened one night with a rather annoying pain in his left shoulder and chest. It was of the gnawing, neuralgic type with nightly exacerbations. At the end of a week, the pain involved the whole arm and the left sciatic distribution. There was no temperature at any time nor did the indisposition force the patient to take to his bed. A neurologist who was consulted found pupillary anomalies which he considered luetic, and he advised a series of salvarsan injections although the blood Wassermann was negative. These promptly increased the pains and initiated the development of a luetic complex of marked severity. This state of affairs continued for four months with additional symptoms of persistent insomnia and rapid loss of weight (thirty pounds) with profound depression. He continued at his work, however, and was only forced to give up from sheer exhaustion incidental to the pains, insomnia and severe nutritional disturbance. Another symptom which annoyed the patient considerably was a frequent and urgent desire to empty his bladder. This continued practically through a period of four months and was most distressing during the night.

Examination.—The neurological examination revealed as the only positive findings a marked deep hyper-reflexia, zones of hyperesthesia, diffuse muscular tenderness and pupils which showed slight inequality, irregularity in one pupil with slowed light and consensual reaction. Lumbar puncture revealed the fluid as follows:

Pressure	normal
Cells	25 per cm.
Globulin	faintest possible trace
Albumin	two plus
Sugar reduction	increased
Wassermann	negative in 2.0 c.c.
Vernes reaction	negative, tint 8
Colloidal Gold	0112210000

Under complete rest, forced feeding and tonic treatment rapid recovery ensued. The neuralgic phenomena while still present are very mild in character and in no way interfere with the patient's activities. The luetic complex promptly disappeared when the nature of the illness was explained to the patient.

Case III. L.S., a married woman thirty years of age, whose previous history was negative, suddenly developed a severe burning sensation across the lumbar area which radiated down the posterior aspect of both thighs. This was accompanied by severe boring and darting pains in the distribution of both sciatic nerves. A sense of apprehension, irritability, insomnia, scalp pains and marked fatigue completed the clinical picture. A slight rise of temperature was present for two days thereafter the temperature continued normal for a period of three months during which time the patient was under continuous observation.

Examination.—The neurological examination revealed as the only positive findings, marked muscular sensitiveness, areas of hyperesthesia limited to the lumbosacral distribution and hyper-active deep reflexes. Lumbar puncture revealed the fluid as follows:

Pressure	considerably increased
Cells	12 per cu. mm.
Globulin	absent
Sugar reduction	increased
Wassermann	negative in 2.0 c.c.
Colloidal Gold	0011110000

Under hydrotherapy, tonics and a trip to the country there was considerable improvement in the symptoms, although the patient complains bitterly of persistent muscular soreness and diffuse neuralgic pains.

Case IV. F.L., male, age fifty-four, salesman from the service of Dr. Henry W. Frauenthal at the Hospital for Joint Diseases, whose previous history apparently had no bearing on his present complaint, developed rather suddenly over the sacro-iliac joint a severe pain which radiated down the outer aspect of his left thigh and leg. This persisted for several days and finally disappeared, to be replaced by a dull ache over an area sharply limited to the upper third of the outer aspect of the leg. There were no paresthesias or other abnormal sensations. The patient did not appear ill nor were there disturbances in the pulse or temperature. The neurological examination revealed as the only abnormalities an absent left supra-patellar and patellar knee-jerk, and weak abdominal reflexes. There were no pupillary anomalies. A careful sensory examination revealed no obvious changes. Lumbar puncture showed the fluid as follows:

Pressure	under moderate tension
Cells	15 per cu. mm.
Globulin	absent
Wassermann	negative in 2.0 c.c.
Colloidal Gold reaction	not done

His condition remained unchanged during the period of observation.

Case V.—This patient was a married woman thirty years of age. Her previous medical history aside from the usual disturbances incidental to motherhood presented no abnormal features. About a year previous her husband had deserted her, leaving her and a child practically destitute. As a result of this unexpected and unfortunate turn in her marital relation she became markedly nervous and apprehensive, was greatly depressed and with great difficulty tried to adjust herself to her misfortune. She slept poorly, worried a great deal and had periods of marked emotional disturbances. This state of affairs continued with varying degrees of severity for a period of six or seven months. At the end of this time she developed rather suddenly lancinating, gnawing pains in both upper and lower extremities, in the chest and lower spine. This condition had persisted three months with slight remissions when she appeared in the Clinic of the Hospital for Joint Diseases asking for relief. In addition to the above complaints there were marked emotional instability and an overpowering sense of fatigue. The examination revealed no disturbance in pulse or temperature. The entire bodily musculature was markedly sensitive to pressure with areas of hyperesthesia that were distinctly segmental in distribution. The pupils were unequal, there was definite irregularity in their contour and one showed a sluggish light reaction. The deep and superficial reflexes showed no variation from the normal. The lumbar puncture showed the fluid as follows:

Pressure	considerably increased
Cells	22 per cu. mm.
Globulin	faint trace
Sugar reduction	increased
Wassermann	negative in 2.0 c.c.
Colloidal Gold	0121110000

The lumbar puncture was followed by a marked increase in the pains, vertigo and severe bitemporal headache.

Case VI. N. L., a business man thirty-eight years of age. The antecedent history in brief is as follows:

July, 1919, a non-confining illness characterized by lassitude, irritability, loss of weight and apathy. Apparent recovery after three months' rest at a northern summer resort. Upon the resumption of business the lassitude and irritability returned.

April, 1920, a recurrence of the symptoms enumerated above with the addition of marked anxiety. Physical examination at this time revealed tachycardia, exophthalmos, tremor of the fingers, dermographia and severe peridental infection with many root abscesses. The diagnosis of psychoneurosis (anxiety type) and hyperthyroidism predicated upon focal infection of the teeth was made. Radical dental surgery, which necessitated hospital confinement, was followed by marked subjective and objective improvement that continued for two months.

August, 1920, a sudden onset of headache, sense of fullness in the stomach after meals with gaseous eructations, intestinal distention, and pain in the scalp. There was a febrile period of twelve days during which the temperature averaged 100° F. The lungs were negative to physical, fluoroscopic and roentgenologic examinations. Apparent recovery was followed by persistence of the irritability and marked diminution of physical vigor.

January 24, 1921, while *en route* to a southern winter resort there was a sudden onset of abdominal discomfort with eructation of gas after meals, intestinal distention, obstinate constipation, soreness and deep-seated dull pain in the lumbar and abdominal muscles, frequent micturition and pain in the scalp. The temperature averaged from 99° to 100.3° F. with a few remissions for one month and anorexia became extreme.

February 23, 1921, the patient was seen in consultation by Drs. M. W. Marr, F. E. Jones, Harlow Brooks and L. H. Cornwall at Pinehurst, N.C. Examination revealed the following positive neurologic findings: hyperactive deep reflexes, diminished upper left lateral abdominal, bilateral Babinski, Chaddock, Oppenheim and Schäfer, left transient ankle clonus, left homonymous pallor of the optic discs with some thickening of the retinal vessels, dermographia, Sargent's line. The lumbar puncture showed the fluid as follows:

Pressure	moderately increased
Cells	185 per cu. mm., polymorphonuclears, 34%, lymphocytes 66%
Globulin	two plus
Sugar reduction	increased
Albumin	increased
Wassermann	negative in 2.0 c.c.
Vernes reaction	negative in 1.6 c.c.
Colloidal gold	2222345321

With rest, nourishing food and symptomatic medication the symptoms gradually subsided. The last to disappear was the dull intermittent pain in the lumbar region which was referred around the body like a girdle. By the end of March all of the symptoms had disappeared except the pain, which was less constant and of diminished intensity, and the left Babinski. At this time there was also noted a narrow girdle of hyperesthesia to cotton wool and hyperalgesia to pin-prick encircling the body just below the costal margin.

Lues was considered as the etiologic factor in this case, but the absence of any history of infection or of previous symptoms, the negative marital history, the sudden onset with gastro-intestinal symptoms and hyperthermia, the spinal symptoms, the absence of pupillary disorders, the negative blood and spinal fluid serology with the other positive spinal fluid findings, were considered sufficient to exclude this, and the diagnosis of encephalitis or encephalo-myelitis was made.

COMMENT ON THE CLINICAL ASPECTS

In presenting the histories and the clinical and laboratory findings of the six cases emembodied in this paper, we feel warranted in considering them as atypical forms of epidemic encephalitis. While not unmindful of the fact that a plethora of types has been described in this affection, the peculiarity and incompleteness of the clinical picture justifies our contention in giving these cases special recognition. It is quite obvious that the major symptoms that we are wont to associate with epidemic encephalitis are entirely lacking in the cases herein described, the diagnosis being made possible only by careful correlation of the clinical and laboratory data. A striking feature of these patients was the fact that with the exception of Case I, they were all ambulant and were with difficulty persuaded to go to bed. This is surely atypical when we consider that the average patient with encephalitis is forced to bed quite early in the disease and stays there for weeks or months. Cases II and V, in view of the abnormal pupillary phenomena, were certainly suggestive in a clinical sense of neuro-lues, and one of these, Case II, was subjected to an intensive course of salvarsan therapy. The negative Wassermann in the blood and spinal fluid in all of the cases obviously excluded luetic infection.

Case VI was a striking clinical demonstration of the long gaps that occur between the development of new symptoms. This we may assume to be more than a mere recrudescence, as an entirely new clinical picture presented itself each time. This peculiarity in the behavior of epidemic encephalitis has been emphasized by Tilney and others.

Insomnia, emotional instability, absent or very slight rise in temperature, the predominance of purely spinal phenomena such as intermittent dull boring pain of a vague shifting character, muscular sensitiveness, scalp pain, bladder disturbances, plegias, alterations in reflex behavior, were other marked atypical features distinguishing these cases from the classic lethargic type of epidemic encephalitis.

In the light of our experience, one may safely predicate the existence of many irregular or atypical forms of the affection. These may be purely ambulant in type. The diagnosis can be made with a fair degree of probability from the clinical facts. A study of the spinal fluid is essential, however, for a proper identification of these types.

COMMENT ON THE SEROLOGICAL ASPECT

An increase in the pressure was a constant finding, which we take to be only an indication of some form of irritation.

The cell counts were all pathological. This cannot be said to be a constant finding in encephalitis, however, as in typical cases of the lethargic type we have encountered normal (1-5) and borderline (6-9) counts. In Case VI there was an unusually high percentage of polymorphonuclears. The 34 per cent noted in this case was higher than any reported in the series of fifty cases of anterior poliomyelitis cited by Larkin and Cornwall in 1918. In our opinion a comparative study of the differential cell counts in a large series of these two diseases would be an important contribution to the literature of the subject, and might add valuable information for differential diagnosis. In the opinion of one of us (Cornwall) the polymorphonuclear percentage in encephalitis is generally higher than in anterior poliomyelitis.

The globulin content was increased in four of the six cases. As a rule, the globulin is slightly increased in encephalitis.

The sugar content, estimated by qualitative examination, was increased in four cases, normal in one, and not recorded in one. We regret that routine quantitative estimations were not made, as we consider this of definite differential value in diagnosis. In Case II, the qualitative increase was very striking. An increased sugar content in the spinal fluid in encephalitis was first noted by Weil in 1918. He called it non-diabetic hyperglycorachia and stated that it may be present in affections of the peripheral motor or sensory neurones, concussion, ordinary epilepsy, and Jacksonian epilepsy. He attributes a slight increase to the associated congestion, but a pronounced increase to irritation of the center in the floor of the fourth ventricle. In the septic meningitides it is increased early but decreased during the florid stage of the bacterial invasion. Recently Foster has reported hyperglycorachia in nine of ten cases of encephalitis (90 per cent) and emphasizes its value as a diagnostic aid in the absence of pleocytosis, increased protein or abnormal colloidal gold reaction.

The colloidal gold curves in encephalitic fluids are usually low, although one of our cases gave a result on the borderline between Zones II and III resembling the so-called "meningitic" curve. It should be emphasized that the colloidal gold reaction does not serve to diagnose any neurologic disease. Its value lies in its correlation with other laboratory and clinical findings. Results similar to those reported in encephalitis have been reported by Larkin and Cornwall in neurosyphilis, anterior poliomyelitis, low-grade meningitis, epilepsy, alcoholic neuritis, multiple and lateral sclerosis, brain tumor, hemiplegia, chronic nephritis and eclampsia.

The Wassermann and Vernes reactions are negative, with the larger amounts of spinal fluid, unless syphilis be coexistent. In the latter case differential diagnosis is rendered very difficult but the quantitative sugar content may determine the diagnosis.

SUMMARY

The occurrence of the cases described at the end of a period during which encephalitis was present in epidemic form, suggests that the same etiologic factor is concerned, but in attenuated form, producing milder symptoms and less typical clinical pictures. In a descriptive sense the term encephalitis does not seem strictly appropriate, as the symptoms point to essential lesions in the cord rather than the encephalon. Encephalomyelitis or meningo-myelitis are terms better adapted to these cases than is encephalitis.

In every case in this series a lumbar puncture was necessary for confirmatory diagnosis. These ambulatory cases may be easily mistaken for neurosyphilis, ganglionitis, neuritis, myositis or psychoneurosis. In our opinion they are more frequent than generally believed, but are overlooked because of the paucity of the symptoms.

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A CASE OF OPTIC ATROPHY*

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This case is presented before the conference because:

1. It illustrates the necessity of a complete serological examination in any given neurological condition.

2. It demonstrates one of the various ways in which neurosyphilis may manifest itself clinically.

3. It is hoped that this case will arouse discussion on the different etiological possibilities that might cause such clinical symptoms, and will help to clarify some of the perplexing factors encountered in such cases.

M. H. (31,529) Male. Forty years old. American. Single. Clerk in hardware store. Admitted March 17, 1921, with a letter from the out-patient department of the Knapp Memorial Hospital, stating that the patient was suffering from bilateral optic atrophy, and asking for possible neurological etiological factors responsible for the condition.

History.—Information was given by the patient himself, who appeared to be of a free and somewhat indifferent disposition. He stated that on February 22, 1921, when he awoke from his sleep, he saw double. He went to work, where he was told by a fellow worker that his left eye was "turned out." He finished his day's work, but did not return on the following day because of nausea, dizziness and general malaise. He then noticed that his right eyelid was drooping, and that his vision was becoming poor. The diplopia persisted for two weeks, and then disappeared. He went to the Knapp Memorial Hospital Out-patient Department, and received glasses. His vision began to fail on March 1, 1921, and was worse in the right eye. The glasses improved the vision in the left eye, but failed to do so in the right eye. He could not sleep.

His previous history was uneventful. He knew nothing of his birth and of his infancy and childhood. He began to go to school at eight years, but was always playing truant. He had never received any head injuries. He denied gonorrhea and syphilis, both by name and by symptoms. He could not recall any former illness. He described his habits as being temperate.

His family history was negative for any chronic or nervous diseases.

Physical Examination.—The patient came into the examination room showing evident ptosis of the right eyelid. His gait was normal in all phases. His station was steady. There was no Romberg. The non-equilibratory tests were all performed normally. There was no dysmetria, no dyssynergia and no adiadochokinesis. Skilled acts were performed normally. Speech did not reveal any defect and the test phrases were spoken very well. There were no abnormal involuntary movements seen.

Reflexes:

	LEFT	RIGHT
Jaw	0	0
Pectoral	0	0
Biceps	1	3
Triceps	1	3
Radial	1	3
Ulnar	1	3
Wrist	1	2
Suprapatellar	3	3 plus
Patellar	3	3 plus
Achilles	1	2
Ankle clonus	0	0
Abdominal	1	1
Cremasteric	1	?
Babinski and modifications	0	0

Muscle strength was well preserved.

Muscle tone was definitely increased in the right extremities.

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*Presented before the Conference of the Neurological Staff of Columbia University at the Vanderbilt Clinic, on April 4, 1921.

There were no abnormal associated movements.

The general sensory examination revealed a normal response to all stimuli.

CRANIAL NERVES showed the following:

Olfactory nerve and nose showed nothing abnormal.

Visual acuity: Left equals 20/80; right equals 20/80.

Visual fields were concentrically diminished.

The fundi showed bilateral optic atrophy more advanced in the left. The pupils were equal, somewhat irregular in outline, but both reacted well to light and accommodation. There was ptosis of the right eyelid. There was definite nystagmus, on looking to the right, greater in the right than in the left eye. The left palpebral fissure was wider than the right.

Acoustic nerve and ear were normal.

Trigeminus nerve and mouth showed nothing unusual. All of the other cranial nerves were found intact.

MENTAL STATUS.—The mental status of the patient was ominous in that the patient showed an attitude of levity and silliness that suggested strongly an exalted and boastful condition, frequently encountered in the parenchymatous cerebral neurosyphilis cases. His intelligence was definitely below par. His emotional response was hardly adequate and hardly consistent with the stimulus, and his mood was quite elevated. His attention was difficult to hold. Memory was intact.

His systemic examination was negative. His skin was warm and of a soft consistency. There was a normal fat distribution. There was no dermographia.

The glandular system was normal.

His heart was normal in outline, of normal force and rhythm, and showed a harsh systolic murmur at the apex. Pulse was 84. Blood pressure 135/85.

Lungs were of normal resonance, breath and voice sounds.

Gastro-intestinal tract was negative.

Genitals were normal.

Blood Wassermann, March 17, 1921, 4 plus.

Spinal fluid showed moderate pressure, trace of globulin, 5 cells. Four plus Wassermann reaction to 2 c.c. of spinal fluid, and collodial gold curve 4442221110.

SUMMARY.—Male. Aged forty years. Single. Clerk in hardware store, of negative family history, who denied any previous illness, who admitted exposure to but denied contraction of venereal diseases, and who was referred by the physicians of a large eye hospital for a neurological examination in an effort to account for a bilateral optic atrophy.

The disease was of three weeks' duration and manifested itself by nausea, dizziness, malaise, diplopia, external strabismus of the left eye, bilateral ptosis and diminishing vision.

On examination the patient presented a superficial, boastful and expansive attitude and an emotionally indifferent and careless mental state, but with a fairly intact memory and orientation. Physically he showed ptosis of the right eyelid, unequal deep reflexes more active on the right side, without any pathological reflexes, increased muscle tone in the right extremities, bilateral diminution of visual acuity, bilateral optic atrophy, more advanced in the right eye, irregular pupils, ptosis of the right eyelid, 4 plus blood Wassermann, 4 plus spinal fluid Wassermann, 5 cells in the spinal fluid and a trace of globulin, and a colloidal gold curve of 4442221110.

COMMENT ON CASE.—The diagnostic possibilities presented in the inverse order of probability are:

1. Accessory sinus disease.
2. Epidemic encephalitis.
3. Neurosyphilis.

1. That accessory sinus disease plays an important rôle in many cases of impaired vision has been forcibly brought to the attention of medical men by the work of White[1] and other rhinologists, although this casual

relationship, especially in its bearing on choked disc, has been questioned by Cushing.[2] In the case presented, we do not think there is any evidence of accessory sinus disease. It is mentioned simply to note that this possibility was not overlooked.

2. Epidemic encephalitis must be regarded as a strong possibility, in view of the presenting symptoms. The general malaise, diplopia, ptosis of one eyelid and nystagmus, appearing during an epidemic, must make one think of this disease which is so protean in its manifestations.

3. Neurosyphilis is, in our opinion, the disease in this case. The strongly positive blood Wassermann leads one to believe that not alone is the nervous system involved in the pathological process, but that the disease is systemic as well. The colloidal gold curve would point towards a strong parenchymatous involvement. The euphoria and inadequate emotional reaction would point towards a cerebral involvement. These two facts would make one think of the possibility of an impending paretic condition, and would make the ultimate outcome of the disease rather unfavorable.

In our opinion, this case should immediately be put on intensive intravenous arsphenamin administration combined with intramuscular mercurial injections. We do not think that at present there is any indication for intraspinous therapy. The case should be carefully followed up and should never be allowed to get away from medical care and supervision.

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DISCUSSION

Dr. Osnato. I would like to have Dr. Cornwall express an opinion concerning the prognosis following treatment. In those patients whom we have in the clinic, I have seen no improvement at all. We have had many early cases, and it is possible that the progress in these has been stopped. I should like to know what Dr. Cornwall would promise a patient with this condition concerning his sight.

Dr. Cornwall. I would not promise him anything. I would tell him that his sight might improve, but I would not think so myself.

Dr. Aronson. Dr. Sands has proven that the patient has two things: lues and encephalitis. But that the lues is the cause of the optic atrophy is not proven. This man might have had lues for some time. The suddenness of the atrophy would perhaps suggest its being due to the encephalitis rather than to the lues. His symptoms may be due to syphilis, but I do not think it can be proven that they are not due to encephalitis.

Dr. McKendree. I understand that this patient was sent here with optic atrophy. I would like to ask whether anyone has seen optic atrophy with epidemic encephalitis. We have all seen cases with papilledema, but has anyone seen a real atrophy in epidemic encephalitis?

Dr. Osnato. May I ask Dr. Cornwall whether he would consider the presence of an optic atrophy in a luetic patient a contraindication to treatment by salvarsan?

Dr. Cornwall. No, I would not consider salvarsan treatment contraindicated.

Dr. Aronson. Answering Dr. McKendree's question, I would like to say that I have a brain which I will soon be able to present at the conference. The patient had encephalitis, his first symptom being a tic involving the 5th nerve. On the first morning there was a ptosis and the patient was blind in one eye. He became unconscious and it was difficult to make an examination, but prior to his becoming unconscious a choked disc was made out and he claimed that he could not see at all. He then became unconscious and died soon afterward.

Dr. Tilney. I am very much interested in the case and have not much to add except that from now on, I presume, we are going to be in a state of combat as to the simultaneous coexistence of syphilis and epidemic encephalitis. I can see no reason why the two might not coexist. Yet it does seem to me, from what

I have seen and what I know of the literature, that syphilis by itself is capable of producing all of the symptoms which we have witnessed in this patient, and that unless the evidence of the coexistence of other disease is unusually strong and convincing, we are going out of our way to invoke this popular subject of encephalitis. I believe that as the case stands everything about it may be explained on the basis of syphilis. In the first place, the suddenness of the onset does not exclude syphilis. Syphilis does begin in many different manners, develop in many forms, and yet preserve its "unity in variety." Here the question of optic atrophy seems to stand in the way of believing that it is epidemic encephalitis. I myself would much prefer to believe that the optic atrophy is due to luetic optic neuritis. So far as the rest of the midbrain symptoms go, an endarteritis or a gummatosis would be sufficient to explain the situation. Until some very telling points are put forth by the proponents of encephalitis, I should consider the case one of neurosyphilis.

DR. POLON. The man has a prominent lower jaw and features that have been pointed out as indicating a special susceptibility to lues. This susceptibility is somewhere related to the perverted function of the pituitary. Oppenheim has described a certain type of tabes which he called pituitary tabes, in which the basal meningitis involved the pituitary. Ordinarily this type was described as non-luetic, but now no such distinction is made, and the term of dyspituitary tabes is used.

DR. WECHSLER. Oppenheim describes in that article two cases diagnosed as paresis and two diagnosed as tabes. All came to autopsy and none showed any relation to tabes. They did show pituitary disturbance. In all these cases one could not make a diagnosis other than paresis or tabes.

DR. CORNWALL. Dr. Osnato asked regarding improvement, which reminds me of the work Dr. Behrens is doing. He has been following these cases for four or five years and has kept records of them. In some of the more recent ones we have just been using drainage alone, and I would like to ask Dr. Tilney if there is reason to believe that this treatment would cause improvement. I have considered atrophy to mean a scar due to actual degeneration of the fibers. How much may we expect those fibers to regenerate in lues?

DR. TILNEY. We must not forget that the optic nerve is not a nerve at all—it is a tract. It contains none of the potentialities of regeneration and recuperation common to peripheral nerves. When an atrophy is once established it is permanent. Those fibers which actually become atrophied do not return to normal. So that the possibility of producing improvement by any means of treatment—intravenous, intraspinous, intracerebral, or anything else for that matter—is quite out of the question. You may arrest further atrophy, but you can never produce any improvement in an actually existing atrophy.

DR. OSNATO. The article by Oppenheim quoted by Dr. Polon and Dr. Wechsler has made me feel that it is something that might better have never been written. How can tabes depend on anything that can happen to the pituitary gland?

DR. VERMILYE. I treated some cases of optic atrophy for Dr. Swift, and I remember one case that we treated over a course of ten years. At first the patient had no vision for anything except light. Within six months he was able to read the headlines in the newspapers. It was a patchy sort of atrophy, and as he recovered his vision, he saw as though he were looking through a sieve.

DR. WECHSLER. I believe Dr. Tilney may recall the case of Mrs. Simmons. A diagnosis of multiple sclerosis was made at first, but after three months it was decided that it was not multiple sclerosis at all. This woman had a bilateral complete optic atrophy. She would lose her vision completely for a couple of weeks and then begin to see again. That repeated itself several times, first in one eye, then in the other. During her stay in the hospital her vision had several times disappeared completely and several times returned so that she could see her way around the ward. She had other symptoms which were puzzling. In the periods during which she could see her fields were contracted.

DR. CASAMAJOR. This discussion shows exactly the position that optic atrophy holds today, the extreme ideas that all of us have about it based upon an insufficient knowledge of the pathology. For instance, we have been seeing some unusual cases in the past few weeks. Some may have been due to wood alcohol poisoning, but not all. And they seem to get better. They come on rather suddenly and get worse for a while, and then for some unknown reason improve. They are not luetic and not wood alcohol, and I think they may be encephalitic. Dr. Aronson's point is well taken. Even if this patient has syphilis, the optic atrophy may be due to encephalitis. The encephalitis that we have seen in the past year or two has varied considerably, and I am not at all certain that these cases may not be encephalitis, especially when judged from their course and their getting well without treatment. In my experience this is a thing which does not happen in luetic atrophy. I think we shall have to revise our notion of optic atrophy. The optic nerve is a tract, as Dr. Tilney says. The cells lie out in the periphery. Besides this, the degenerations are not descending. Pressure anywhere along the course of the nerve will cause degeneration back to the cell itself. The ascending degeneration from a point of injury, up to the nerve, is a reaction which is different here from other structures of the nervous system. Then as to the possibility of recovery there is another problem. Obviously there could not have been an atrophy—a real degeneration of all the myelin sheaths with the later disappearance of the axones and the replacement of that tissue by neuroglia. There must have been some other kind of pathological process. We call it atrophy, but I wonder if some other condition may not also cause this degeneration. I think the pathology of the optic nerve is an almost untouched field. We do not know enough about the anatomy of the retina. We certainly know nothing of the pathology of the ganglionic layer of the retina, and we know even less of the pathology of the optic nerve itself.

DR. ELSBERG. Does not this apparent recovery of vision in optic atrophy mean that what is reported as complete optic atrophy by the ophthalmologist is not really a *complete* optic atrophy?

DR. SANDS. I have never seen atrophy in epidemic encephalitis, although I have seen much choking of the discs. I have seen four patients with choking of the disc to 2½, 3 and 4 diopters, with hemiplegia on the opposite side, and conditions so closely resembling brain tumor that the only thing which stood in the way of operation in one case was getting the consent of the family. Now that patient is getting well without operation. I have seen one case which recovered from the condition and now shows a haziness of the disc but not a real atrophy. He had a choking of 4 diopters of the discs.

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MENINGOVASCULAR NEUROSYPHILIS*

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PART I

DEFINITION.—Meningovascular neurosyphilis is that form of luetic infection which expresses itself pathologically as an involvement of the meninges and blood-vessels of the nervous system. Primarily it produces changes in the mesodermal elements which participate in the formation of the cerebrospinal axis. Secondarily, it may compromise the actual neural constituents, including the nerve cells and nerve fibers. In this respect meningovascular neurosyphilis differs from paresis, taboparesis, and tabes dorsalis, all of which latter result from the direct attack of the infecting organism upon the neural components.

That there is an absolute boundary line between these pathological forms of neurosyphilis cannot be conclusively maintained at the present time. On the other hand, for the purpose of classification and clinical distinction, it is convenient to recognize the differences just described. In many cases the clinical and laboratory findings will not suffice to establish the distinction with a clarity that might be desired; and this fact seems to justify the statement of Southard that "no one can now successfully make a differential diagnosis between the paretic and the diffuse nonparetic forms of neurosyphilis in many phases of either disease, even with all laboratory refinements."

HISTORICAL NOTE AND NOMENCLATURE

Much conflicting evidence exists concerning the early recognition of syphilis. Whether it was a totally new malady which swept over Europe in the latter part of the fifteenth century, or whether it was an epidemic recrudescence of a disease of much greater antiquity, is still a matter of doubt and debate. It seems certain, however, that this infection made a virulent appearance in Europe during the Middle Ages, at a time when humanity was plagued by epidemic diseases probably as never before or since. The interpretations given these widespread pestilences naively laid the responsibility for their existence to adverse astral influences, to famines and failures of crops, to volcanic eruptions, inundations and droughts, to the pollution of wells by the Jews, to plagues by insects, and to other equally irrelevant causes. The actual and cogent factors predisposing to medieval pestilence were the prevailing ignorance of the times, the neglect of public and private hygiene, the almost total lack of sanitation of the ancient walled towns, the misgovernment and unbridled immorality growing out of many wars and constant preparation for war, which brought into intimate contact all kinds and conditions of people belonging to many different races.

During this period at least eight epidemic diseases were recognized, namely, the *bubonic plague, phthisis, scabes, erysipelas, leprosy, anthrax, trachoma and syphilis.* Of these, the black death and syphilis were by far the most malignant. The black death,

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after devastating Asia and Africa, appeared in Europe about 1350. During its course this pestilence destroyed twenty-five million Europeans and is credited with the unprecedented mortality of nearly sixty million human beings throughout the world.

Somewhat later in the Middle Ages, while the black death was still prevelant but already giving evidence of attenuation, syphilis appeared as the second great scourge in epidemic form, following the siege of Naples in 1495. By some it is thought to have been communicated to the French invaders by the Spanish, who, it was believed, had become infected by the sailors of Columbus following the discovery of the New World.

Medieval syphilis was first known as *mal franzoso, morbus gallicus* and *mala napoletana*. It was not until 1530, however, that the disease acquired its present name at the hands of the poet-physician, Girolamo Fracastoro, in consequence of what is perhaps the most celebrated of all medical poems, "Syphilis sivi Morbus Gallicus" (Venice, 1530), which describes the disease, takes cognizance of its venereal origin, and assigns to it its present name.

The relation of syphilis to the nervous system, and the possibility that it might affect the central controlling organs of the body, was apparently recognized during this first great epidemic of the disease in Europe. At all events, Leonicino, in 1497, seemed to be aware that syphilis might involve the internal organs and even produce paralysis. Subsequently, Paracelsus and Van Hutten observed involvement of the internal organs, and Van Hutten maintained that in certain instances syphilitic individuals became paralyzed. It is probable that the first actual reference to syphilis of the brain was given by Morgagni in his statement that gummata do not limit themselves exclusively to the periosteum and bones. The quotation indicating this early recognition of cerebral gummata is of much historical importance and will be found in the original appended in a footnote, together with Morgagni's description of aneurysms and diseases of the smaller vessels in the brain observed postmortem in a syphilitic subject.[1] Mention of syphilitic disease producing changes in the functions of the central nervous system is made in 1740 by Astruc in his book *De Morbis Venereis*.

It was unfortunate, perhaps, that the slowly advancing recognition of syphilitic involvement of the nervous system met a setback at the hands of so distinguished a leader of medicine as John Hunter in about 1790. His teaching was that syphilis never affects the internal organs, so that—"It would appear that some parts of the body are much less susceptible of the lues venerea than others; and not only so, but many parts so far as we know are not susceptible of it at all. For we have not yet had every part of the body affected; we have not seen the brain affected, the heart, stomach, liver, kidneys nor other viscera, although such cases are described in authors."

Early in the eighteenth century the term "lues venerea" was applied as a synonym of syphilis. Originally lues was a generic term of much wider application. Its use evidently dates back to Roman times, when it was employed to indicate a plague, pestilence or contagious disease. It is used by Virgil as implying a pest or a reproach for criminals, and by Cicero for any widespread or universal calamity. In relatively modern times only has the term received its present significance in relation to syphilis.

The influence of Hunter's dictum, which apparently retarded the advance of knowledge concerning central nervous syphilis was in part offset by Lallemand who, in 1835, demonstrated a collection of syphilitic brains. Virchow's writings on the pathology of gummatous lesions went far to establish a firmer foundation for the actual involvement of the nervous system by syphilis, and his teachings were followed by many publications on syphilis of the nervous system.

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[1] "Qui in Theatro hoc nostro tria gummata candida, recluso cranio, ostendi vidit ab Antonio Molinetto quae inhaerebant Durae membranae, verum etiam ex Guarinonio scribente, se vidisse Gummata dicta in cerebro concreta. Quam potissimum sedem memoro, quia haec adscribuntur ad eiusdem Observationem in qua tria corpuscula tanquam gummata virentia in viri cerebro conspecta leguntur, qui ex hoc de qua loquimur epilepsiae et convulsionibus obnoxius, denique gravissimo sopore confectus est." Morgagni, J. B. "De sedibus et causis morborum per anatomen indagatis libri quinque." *Epistola Anat. Med.*, lviii, Art. 9. 1761.

"Sed in Tenui meninge arteriarum trunci omnes, omnesque item earum rami, iique praesertim qui plexum Choroidem versus contendunt, multo erant crassiores aequo, et duriores; exsiccatique osseam pluribus in locis naturam ostenderunt." *Ibid. Epistola Anat. Med.*, xxvii, Art. 28.

In 1868 Allbut gave the first description of the microscopic appearance of syphilitic arteries, and in 1874 there appeared an important monograph by Heubner describing syphilitic disease of the cerebral arteries. A number of investigators since then have shown that not only acquired, but—quite as much—hereditary syphilis may affect the central nervous organs and produce pathological changes of the degenerative, arterial or gummatous variety.

In the latter part of the nineteenth century a considerable discussion developed concerning the distinction of the various forms of syphilis affecting the nervous system. It is to Fournier (1886) that we are indebted for the first clear conception that locomotor ataxia and probably paresis differ from other forms of nervous syphilis in the late appearance of the symptoms. For this reason these diseases were considered *postsyphilitic*. To describe such late consequences of syphilitic infection, Fournier devised and employed the term *parasyphilis*. His contention that the tabes dorsalis was a postsyphilitic disease received eminent support from the extensive clinical investigation of Erb, whose statistics of 1,100 cases of tabes appeared to justify the recognition of the condition called parasyphilis. Krafft-Ebing lent the weight of his opinion to the conception that paresis was a postsyphilitic condition when he stated that "general paralysis is a product of syphilization and civilization."

According to Mott, syphilis of the nervous system may be divided into two major groups, namely:

1. Syphilitic diseases resulting in basal meningitis, meningitis of the convexity, cerebrospinal meningitis, arteritis, gummatous tumors and syphilitic diseases of the spinal cord.
2. A group comprising parasyphilis, which includes tabes dorsalis, tabo-optica and dementia paralytica.

A number of authorities, among them Nageotte, Lesser and Herschel, maintain that general paresis and tabes are quaternary syphilis. Möbius refers to tabes and general paresis as *metasyphilis,* by this term implying that, although lues may be the fundamental cause, the disease itself is not directly due to the syphilitic virus.

In 1907 Ravaut and Ponselle, in four cases of hereditary and acquired syphilis, demonstrated the presence of spirocheta pallida in the ependymal cells of the cord and the peri-ependymal neuroglial cells. Syphilitic ependymitis, according to these investigators, seems to be one of the stages in the invasion of the nervous system by spirochetae.

In 1913, Noguchi and Moore reported the results of their examinations for the treponema pallidum in paretic brains. One of these authors succeeded in finding the organism in 12 out of 70 specimens. "The cases," Moore says, "in which the pallida were found showed the classical physical signs. The postmortem findings in the brain in every case were definitely those of general paralysis. Seven were of the cerebral type, five of the tabetic." This important observation goes far to establish the fact that general paresis in particular, and probably tabes also, cannot be regarded as after-effects of syphilitic infection. The presence of the organisms in the brain tissue demonstrates conclusively that these diseases are due to an active syphilitic process; therefore to regard paresis as a postsyphilitic disorder places a wrong interpretation upon the condition which may prejudice our clinical understanding of the disease.

In 1917, Southard and Solomon, in an important contribution to the subject of syphilis of the nervous system, clearly demonstrated the existence of a great number of clinical varieties, all of which they grouped under the general term *Neurosyphilis*. They especially pointed to what they regarded as the "unity in variety of the neurosyphilitic phenomena." It was their opinion that "the unity of these phenomena is confirmed, theoretically, by the common factor of spirochetosis, practically, by the Wassermann reaction, positive in serum or spinal fluid. Almost at this point the unity of phenomena ceases. Neither chronicity nor evidence of mononuclear cell deposits, nor evidence of serious structural damage to the nervous system, nor presence of other positive tests than the Wassermann reaction, nor existence of mental or nervous symptoms or signs, is a common feature of neurosyphilis. Sometimes the nervous system appears to harbor spirochetes in the most cordial manner as guest-friends (*paresis sine paresi*). Again, perhaps as an expression of elaborate processes of immunity, the spirochetes take effect in relatively huge gummata. Sometimes the neurosyphilitic process rises as if by a regular process of siege from spinal nerve-root to spinal nerve-root (tabes dorsalis and diffuse neurosyphilis). Again, the nervous system is taken by storm, as it were (disseminated encephalitis). Very frequently the neurosyphilis is simply an indirect effect of blood-vessel disease. The laws governing the preference of processes to lodge in membranes, vessels and parenchyma, and in all combinations of these, have not been worked out."

The more important contributions in the development of knowledge concerning neurosyphilis are summarized by Southard and Solomon as follows:

1858. VIRCHOW. Presentation of the pathology of gummatosis.
1874. HEUBNER. Demonstration of syphilitic endarteritis.
1891. QUINCKE. Introduction of the technique of lumbar puncture for diagnosis.
1901. RAVAUT, SICARD, NAGEOTTE and WIDAL. Cytodiagnosis in the cerebrospinal fluid.
1903. WIDAL, SICARD and RAVAUT. Changes in the albumin content of the cerebrospinal fluid.
1903. METCHNIKOFF and ROUX. Transmission of syphilis to apes.
1904. ALZHEIMER. Presentation of the histopathology of brain syphilis.
1905. SCHAUDINN and HOFFMANN. The discovery of the spirochete in its relation to syphilis.
1906. WASSERMANN, NEISSER and BRUCK. Serum diagnosis in syphilis.
1908. PLAUT. The application of the Wassermann reaction on the cerebrospinal fluid to the diagnosis of syphilis of the nervous system.
1909. EHRLICH. Preparation of salvarsan for the treatment of syphilis.
1912. SWIFT and ELLIS. The preparation of salvarsanized serum for the intensive treatment of neurosyphilis.
1913. NOGUCHI and MOORE. The discovery of spirochetes in the brain tissue of paretic patients.
1913. LANGE. Application of the colloidal gold solution test in the differential diagnosis of neurosyphilis.

From an analysis of the facts thus far accumulated, it appears that neurosyphilis in all its manifold forms is an active spirochetosis, making itself apparent most prominently in symptoms due to defects in the nervous system, but existing, in all probability, as a general systemic infection as well. The recognition of the several forms of neurosyphilis is of advantage only because it lends convenience to description and at the same time provides a formal classification as a basis for discussion.

ETIOIOGY

The Positive Causative Factor.—Since the original investigations of Schaudinn and Hoffmann, it has been generally recognized that the positive causative factor of syphilis is the spirocheta pallida. Schaudinn gave this name *spirocheta pallida* to the organism found in syphilitic lesions, because of its resemblance to spirochetes in general; but within a year he recognized certain features which he considered distinctive enough to give it a separate classification. He then replaced the generic term *spirocheta* with *treponema*.

Concerning the exact classification of this organism there are still divergent opinions.

especially as to whether it is a bacterium or a protozoon. The systematic affiliation of this family with the protozoa was maintained by Schaudinn. Mott believes that the spirocheta pallida possesses characteristics affiliating it more with protozoon than bacterium. He bases his opinion on the fact that the motile behavior of this spirochete is totally different from that of bacteria. Its invasion of the human body produces clinical symptoms more in keeping with trypanosomiasis than with bacterial infections. The symptoms of the disease have a periodicity unknown in bacteriogenic disorders, while the histological lesions of syphilis of the nervous tissues bear a striking resemblance to the protozoal infections causing African sleeping sickness (Trypanosoma gambiense) and dourine (a disease of horses caused by the Trypanosoma equiperdum).

Furthermore, trypanosomiasis corresponds with syphilis in that it is characterized by a succession of eruptions due to the escape of trypanosomes from the blood-stream into the lymph spaces where they eventually produce a polyadenitis.

Whatever decision may be reached in this matter, it is probable that the best classification of the Spirochaetida or Spirillum organisms is that of Noguchi, who has recognized at least six different genera in this family. Treponema (*trepo,* to twist; *nema,* thread) constitutes one large genus of which there are several species, the specific organism of syphilis being Treponema pallidum.[2]

Line of Progression of the Spirochete from the Initial Lesion Toward the Nervous System.—In addition to the discovery of the Treponema in the skin and mucous membrane of the genital and extra-genital organs during the primary stage, the organism has also been discovered in the capillaries of the skin and the perivascular tissue. Occasionally it is found in the blood, but more frequently it appears in the lymph and lymphatic organs; and its presence here, according to the view of Metchnikoff, is a constant feature of syphilis. From the work of Levaditi and Yamanouchi, it appears that after inoculation in the anthropoid apes, the spirochete remains for some time at the site of injection and there undergoes rapid multiplication. These observers believe that the period of incubation of the disease is not dependent on the existence of an evolutional cycle in the Treponema pallidum, but results from the histological lesions produced by the germination of the organism. The period of incubation comes to an end as soon as the cellular changes in the invaded tissues assure the Treponema the necessary nutritive elements. The causative factor of syphilis, having established a satisfactory entente with the human host at the point of inoculation, proceeds to invade the neighboring lymphatic glands, where it undergoes further multiplication in the lymph spaces, producing an adenitis in its environment in order to effect its still further multiplication and growth. Following the lymph stream, the spirochetes eventually cause a polyadenitis and usually find their way into the thoracic duct, thus producing an infection of the blood-stream and causing a sceondary stage which is manifest in a cutaneous eruption. In many cases meningitis has developed before the primary sore has disappeared, thus indicating the early invasion of the meninges by the spirochete. It seems most probable that the line of conduction generally followed by the spirocheta in its progress toward the nervous system is by way of the lymphatics and the perivascular lymph spaces.

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[2] "*Genus.*—Treponema (Schaudinn, 1905). *Type Organism.*—Treponema pallidum (Schaudinn and Hoffmann, 1905). *Measurements.*—Length, 6 to 14 μ; pointed ends. Diameter 0.25 to 0.3 μ: cylindrical. Spiral amplitude, 1 μ; regular, rigid. Spiral depth, 0.8 to 1 μ; very constant. Waves, one or more slight undulating curves may be present. *Axial Filament.*—Doubtful. The whole seems to consist of a spirally wound axial filament. *Chambered Structure.*—Absent. *Membrane.*—Doubtful; if there is one it must be flexible. *Crista.*—Absent. *Terminal Finely Spiral Filament.*—Present. Easily seen in cultures. *Flagella.*—Absent. *Highly Motile End Portions.*—Absent. *Division.*—Transverse or possibly also longitudinal. *Habitat of Genus.*—Two pathogenic and several harmless parasites. *Other Species.*—*Pertenue, microdentium, macrodentium, mucosum, calligyrum, minutum. Staining Properties of Axial Filament and Cell Membrane.*—Membrane not recognizable. *Staining Properties of Body.*—Stains pink by Giesma's solution. *Trypsin Digestion.*—Resists digestion for many days. *Bile Salts (10 Per Cent).*—Disintegration complete. *Saponin (10 Per Cent).*—Broken up in time.." Noguchi, *J. Exper. M.*, 1918, xxvii, No. 5, 583.

AGE INCIDENCE AND PREVALENCE OF NEUROSYPHILIS

It has long been maintained that syphilis acquired late in life predisposes to disease of the nervous system. Nonne's observations show that a large number of patients who contracted syphilis in their sixth or seventh decades, in a few years became tabetic, although he has observed not a few instances of healthy young adults who became victims of severe nervous syphilis. Of twenty-nine syphilitic brains autopsied by Neumann, the following age incidence was recorded:

Between 1 and 18 years of age	1 case
20 and 29 years of age	3 cases
30 and 39 years of age	8 cases
40 and 49 years of age	4 cases
50 and 59 years of age	6 cases
60 and 69 years of age	3 cases
70 and 79 years of age	2 cases

Nonne's figures with reference to age incidence are as follows:

Between 1 and 10 years of age	3 cases
10 and 20 years of age	2 cases
20 and 30 years of age	25 cases
30 and 40 years of age	75 cases
40 and 50 years of age	55 cases
50 and 60 years of age	23 cases
60 and 70 years of age	2 cases

The proportion of men affected by nervous syphilis is much greater than that of women. This may be due to the fact that men are more exposed to influences injuring the nervous system. In Nonne's group of 185 cases, 125 were males and 60 females. In the group of 155 cases studied by the author, 108 were males and 47 females.

RELATION OF THE INITIAL LESIONS TO THE EARLY SYMPTOMS

Naunyn, on the basis of 254 reported cases of neurosyphilis, came to the conclusion that the first involvement of the nervous system makes itself apparent most frequently in the first year after infection. Forty-eight per cent of all cases of neurosyphilis show their first symptoms within the first three years. From this time on, the frequency seems to decrease from year to year. An extensive and severe case of cerebral syphilis with involvement of the spinal meninges developed two months after the primary infection. Another instance is cited as developing its earliest symptoms of neurosyphilis three months after infection. Nonne reports 2 of his cases with syphilis of the brain which developed four months after infection; and another case, three months after the primary lesion, showed symptoms referable to the nervous system simultaneously with the appearance of a roseola.

Neurosyphilis often develops many years after the initial lesion, and gummatous processes may be so deferred that the appearance of symptoms does not occur until thirty or forty years after the primary lesion. According to Patrick, brain syphilis occurs most frequently during the first year of the infection. Following this in point of frequency, comes the second year, and less frequently the third year. Its occurrence is exceptional after ten years. A review of 185 cases observed by Nonne showed that in 83 the first symptoms appeared within periods varying from one to ten years, while in 47 the development of first symptoms was still later. In 55 cases there was a lack of record in this particular. Nonne concludes from this, that neurosyphilis may develop at any stage of the syphilitic infection, and that the tertiary stage is by no means the favorite.

Of the 155 cases of meningovascular neurosyphilis intensively studied by me, an initial lesion was admitted in 43 per cent. The first symptom appeared as follows:

Immediately after initial lesions, 2 cases	3%
Six months or less after initial lesion, 3 cases,	4½%
Five years or less after initial lesion, 8 cases	12%
Six to ten years after initial lesion, 14 cases	21%
Eleven to twenty years after initial lesion, 30 cases	44½%
Over twenty years after initial lesion, 10 cases	15%

Fildes, Parnell and Maitland, as a result of an examination of 624 syphliitic men in the early stages of the disease, found 18 per cent who gave undoubted signs of abnormal conditions of the central nervous system.

Concerning the length of time after infection at which signs of meningeal involvement may apear in the cerebrospinal fluid, Dujardin maintains that these changes are sometimes manifest after a period of two weeks. He believes that when the spirochetes enter the blood, the meninges react with a slight lymphocytosis, although the skin shows as yet no sign of involvement.

According to Fordyce, "infection of the nervous system probably takes place during the secondary stage. A low-grade meningitis may exist for years or the spirochetes may remain quiescent for years until called into activity."

Kaliski and Strauss state that "cerebrospinal syphilis involving one or all of the structures of the nervous system is a disease essentially of younger individuals and occurs comparatively soon after infection."

McDonagh believes that the organisms reach the nervous system via the blood-vessels. They then involve the meninges (mainly the dura) and nerve tissue proper.

Wile and Stokes are of the opinion that the nervous system may be and probably is frequently involved before there are other evidences of the hematogenous spread of the Treponema pallidum.

S. A. Kinnier Wilson has found that syphilitic meningitis may occur as an acute exacerbation in cases of congenital syphilis. It may develop during the secondary period, soon after the cutaneous exanthem, or even in a pre-roseolar stage. On the other hand, it may be an episode in the tertiary stage, arising in the course of a chronic gummatous syphilis long after infection and sometimes when the lesions in the nervous system have appeared to be latent or quiescent.

It has long been regarded that there are different kinds of virus engendered by syphilis. Certain of these possess a definite specificity for damaging the nervous system. Analogies of this specificity are not far to seek. The proneness of acute anterior poliomyelitis to attack the ventral gray columns, or of lead to involve the motor elements in the craniospinal nerves, or of ergot to affect the dorsal columns of the cord, are well known instances of specific predilection. As evidence of such a predisposition on the part of the virus of syphilis to involve the nervous system, a number of instances may be cited—for example, the tendency toward the development of infantile and juvenile tabes and tabo-paresis in children whose parents have been syphilitic but without syphilitic symptoms. Besides this, many cases of conjugal tabes and tabo-paresis have been reported, in not a few of which the husband has developed general paresis and the wife meningovascular syphilis, or else both have become tabetics or paretics. The fact that there are reported cases in which individuals have become infected from the same source, such as that quoted by Nonne in which three men received infection from the same woman, one of whom developed tabes and the other two paresis, seems to indicate that the influence of heredity and social environment is not of great moment in the development of neurosyphilis. Nonne ificity for the nervous system, is the fact heredity nor family predisposition may be regarded as playing much of a rôle in determining the neural involvement.

As opposed to the view that there is a special form of syphilitic virus with a specificity for the nervous system, is the fact that Whalen has reported a more or less generalized syphilitic involvement coexistent with tabes dorsalis. His postmortem observations of cases disclosed the existence of a specific aortitis and myocarditis together with extensive syphilitic lesions in the viscera and other parts of the body.

It seems probable that neurosyphilis is but part of a generalized systemic infection. This observation is of the utmost importance in regard to the treatment of the individual case. Therapy which is directed solely to neurosyphilis does not take into account the probability of syphilis existing in other organs and parts of the body. Heubner makes this position clear when he propounds the question—"Can one speak of lues nervosa when in the same subject one finds degenerations of tabes or paresis along with gummatous syphilis?" Gummatous disease of the brain and spinal cord obviously does not indicate a specific nerve poison, since gummata may be found in other organs and parts of the body.

The involvement of the nervous system by syphilis seems more likely to depend upon the mode of conduction of the Treponema from the site of infection than upon any specificity of the virus for the nervous tissues. The recognized pathways open to the spirochetes in their invasion of the brain and spinal cord are at least two in number: first, the perivascular lymph spaces, and second, the endoneural lymph spaces in the peripheral nerves. Ehrmann has shown, in cases which were studied microscopically in an effort to determine the probable pathway of the organism from the site of the initial lesion in the prepuce, that the spirochete may follow along the course ofthe nerves. In 2 cases in which the prepuce was extensively ulcerated, a number of nerves of the epidermis and dermis contained many spirochetes. These organisms were found not only in the connective tissue of the nerve fasciculus and in the lymph spaces of the nerve sheath, but also in the nerve fasciculus itself and in the nerve fibers. The Treponema lay entangled in these spaces in regular masses and clumps surrounding the nerve bundles. Deeper in the nerve they were imbedded directly between the nerve fibers. Apparently there was no penetration of the medullary sheaths. The axis cylinders were intact and there was no sign of inflammation in the nerve fasciculus. Upon the assumption that the nerves serve as a pathway of conduction to the nervous system, Ehrmann explains the origin of tabes which most frequently begins in the lowest part of the spinal cord. He is also of the opinion that the transit of the Treponema through the nerves may be slow, and this may account for the long period of latency in the ultimate appearance in the nervous system.

FACTORS INFLUENCING THE SEVERITY AND COURSE OF NEUROSYPHILIS

The severity of the neural involvement in consequence of syphilis has, by many authorities, been regarded as having a definite relation to the primary infection. Cases with mild primary and secondary stages are prone to develop neurosyphilis. This view was first advanced by Broadbent and later supported by Erb and Fournier. Nonne, however, is inclined to question this apparent relation between the severity of the initial lesion and the involvement of the nervous system. A number of cases of brain syphilis ran a severe course from the beginning of the infection, the primary lesion assumed a malignant character, the eruption was papular and pustulo-papular, with new exacerbations during the antispecific treatment.

Brain syphilis has been observed by Oppenheim following both mild and severe primary and secondary lesions.

Although it has been maintained that extra-genital infection bears special relation to the development of brain syphilis, many cases on record fail to substantiate this supposition. Hahn has reported 307 cases of extra-genital infection, none of which developed syphilis of the brain.

Injuries of the head have been considered to exert an influence upon the development of brain syphilis, causing a latent process to take on activity, and in many cases to eventuate in a fulminating neurosyphilis. Not a few isolated instances which seem to substantiate this relation of trauma, especially to the head, have been reported from time to time, yet in the mass of collected statistical studies the influence of injury to the head seems to assume a rôle of minor importance. Nonne, for example, had an opportunity to observe yearly 150 cases of severe injuries to the head in his hospital practice. Among these there was a fair percentage of individuals who had acquired syphilis. He is able to recall but 2 cases in which symptoms of cerebral lues developed after trauma.

According to Bailey, although it seems fair to presume that trauma may produce a *locus minoris resistentiae,* nevertheless, syphilis of the brain following head injuries occurs with surprising frequency.

A special rôle has been attributed to mental strain in the determination of syphilis of the brain. It has been maintained that the man who does manual labor is less likely to develop brain syphilis than the man who is a brain worker. This contention, however, has been disputed. Tarnowsky in 100 cases of cerebral lues found but 6 in which a contributory factor seemed to be cerebral overactivity. Similarly, psychic trauma and acute infectious diseases are regarded as having but insignificant influences as predisposing or contributory causes.

That the long continued or excessive use of alcohol predisposes the syphilitic patient to the development of neurosyphilis, is a familiar teaching. It is certainly true that cerebral syphilis and chronic alcoholism often coexist. Lochte believes that syphilis in alcoholics is most often of the severest type. Tarnowsky in 100 cases of cerebral syphilis found 45 who were habitual drunkards. Nonne points out, however, that the organ most often involved by syphilis in alcoholism is the liver and not the brain.

Influence of Psychopathic and Neuropathic Heredity.—While the inherent weakness due to a congenitally underdeveloped nervous system has been presumed to have a direct influence as a predisposing cause in cerebral lues, and in spite of the fact that several authorities consider a poor nervous inheritance as predisposing to the development of cerebral lues, Nonne's figures in 185 cases of cerebrospinal and spinal syphilis showed that there was no history concerning heredity in 24 cases, a good family history in 136, and a poor family history in 25. This would seem to indicate that the influence of psychopathic and neuropathic heredity has but little bearing upon the predisposition of syphilis for the nervous system.

Influence of Antispecific Treatment.—Another important factor which has long been regarded as having a definite bearing upon the development of syphilis of the brain and spinal cord is antispecific treatment administered in the primary and secondary stages of the infection. It is a question of much practical importance as to what value thorough or insufficient antispecific treatment may have during the early phases of the disease in preventing an extension to the nervous system. Collins, from a study of 96 cases,concludes that thorough treatment of systemic syphilis does not prevent nor postpone the development of neurosyphilis. In Nonne's experience, cases which received energetic antispecific treatment immediately after the infection and at every secondary outbreak, are those which develop cerebral syphilis within the second or third year after the initial lesion. Schuster, reporting 76 cases of admitted cerebrospinal lues, found that 10 were never treated (13.2 per cent); 36 had one course of antispecific treatment (47.4 per cent); 15 had two courses (19.7 per cent); and 15 had from three to six courses of antispecific treatment. On the basis of these figures Schuster was unable to distinguish perceptible differences in the clinical course in the cases which had received previous treatment and those which had not. His analysis also shows that the average period of latency for treated cases was 12.2 years, slightly longer than the average period of latency for untreated cases. In this regard Schuster's observations coincide with those of Collins.

In spite of this evident difference of opinion as to the inhibitory influence of treatment during the initial stages upon the development of neurosyphilis, all authorities are agreed that the apparent failure of early treatment to inhibit neurosyphilis should in no way influence the administration of antispecific treatment in the early stages of the disease.

PATHOLOGICAL ANATOMY AND PATHOLOGICAL VARIETIES OF MENINGOVASCULAR NEUROSYPHILIS

The essential pathological reaction of meningovascular neurosyphilis is confined primarily to the mesodermogenous tissues entering into the nervous system and its coverings. Such secondary effects as may be produced in the ectodermogenous tissues of the nervous system, including the nerve cells and nerve fibers, are for the most part dependent upon the changes in the blood-vessels and coverings of the neuraxis. To draw a hard and fast line in all cases of neurosyphilis between these two types of pathological alteration is impossible, for the reason that there is so much involvement of both types of tissues entering into the formation of the nervous system. This undoubtedly has justified Southard's recognition of diffuse neurosyphilis, by which term he implied the simultaneous involvement of the elements derived from mesoderm and ectoderm. On the other hand, a study of pathological changes in neurosyphilis makes it clear that the impact of the treponemal invasion may fall with preponderant force upon the blood-vessels or the meninges, and thus justifies the conception of a meningeal and a meningovascular type of syphilis.

The possible combinations in which these changes may exist in the mesodermogenous tissues permit the recognition of certain pathological varieties of meningovascular syphilis. Thus, in certain instances, the arteries and, to some extent, the veins are most affected, in which case the pathological condition is known as *syphilitic endarteritis*. In other instances, the coverings of the brain, particularly upon its base and convexity, or the meninges of the spinal cord, show the principal reactions to the treponemal invasion. Such condition is known as *syphilitic meningitis*. Another variety of pathological alteration observed in many instances resembles in all respects one of the best known manifestations of the disease in other organs, namely the formation of gumma. This new growth may implicate the bone or the coverings of the neuraxis. It may take origin in some deeper portion of the brain. It is the pathological variety known as *gumma* or *syphiloma*. When the meninges and arteries are simultaneously affected, as occurs not infrequently, the variety of neurosyphilis is known as *syphilitic meningo-endarteritis* which may be accompanied by thrombosis of the cerebral or spinal vessels, thus giving rise either to encephalomalacia or myelomalacia. Another pathological variety, perhaps of less frequent occurrence than any of those already mentioned, is *syphilitic meningo-encephalitis* in which the pathological changes in the brain result in a diffuse and extensive reaction throughout the cerebral substance as well as in involvement of the arteries.

The pathological varieties, therefore, of meningovascular neurosyphilis include *syphilitic endarteritis, syphilitic meningitis, gumma, syphilitic meningo-endarteritis* with encephalomalacia or myelomalacia, and *syphilitic meningo-encephalitis.*

Syphilitic Endarteritis

In this variety of central nervous syphilis the arteries are extensively involved. There is a thickening of the vessel walls with a narrowing and displacement of their lumina. Their conductivity as vascular channels becomes impaired. The arteries instead of being thin and transparent are firm and cylindrical. They are not easily compressed or flattened. In color they are a yellowish gray, showing this coloration in patchy spots or streaks. Occasionally, grayish nodules appear scattered along the arterial trunks and their principal branches. When such branches are seen in cross section their lumina are almost obliterated by the thickening in their walls. In the smaller arteries this obliteration may be complete. The lumen of the vessel instead of being circular or subcircular, is asymmetrically crescentic or irregularly elliptical. This is particularly true of the basal arteries entering into the circle of Willis. The microscopic examination of such vessels shows a marked increase in cellular elements situated between the endothelium and the fenestrated membrane. The change seems to be essentially an endarteritis which, according to Heubner, starts as a proliferation of the endothelium and extends to the vasa vasorum of the media. It seems more likely, however, that the process starts as a defense reaction by a thickening of the intima in response to the presence of the Treponema in the perivascular lymph spaces. This view gains in probability from the fact that the endothelial cells of the capillaries, arterioles and venules do not undergo pronounced proliferation, whereas the arteries which are surrounded by perivascular lymph spaces show the most marked proclivity toward vascular change. The entire course of the pathological process in the blood-vessels seems to be one of defense against a hostile invasion, during which the efforts at protection often defeat themselves, first by impairing the lumina of the vessels and then by favoring thrombosis. Under such circumstances, many of the vessels, by a process of fibrosis, become converted into more or less solid strands of connective tissue.

Another possibility developing during this change in the arterial wall is the appearance of certain areas of weakness which permit the dilatation of the vessel and result in the rupture of the elastic coat. If the reaction in the media occurs with a sufficient rapidity an aneurysmal dilatation will be formed. Changes in the larger vessels may gradually obliterate the lumina, lead to the formation of thrombi, or, by the development of aneurysms, seriously impair the brain tissue itself. The development of an extensive thrombus deprives the corresponding area of its blood supply so that encephalomalacia or myelomalacia occurs. The rupture of an aneurysm results in hemmorhage which may compromise extensive regions of the brain or spinal cord.

Falling short of these more extreme eventualities due to defective blood supply, the thickening of the arterial wall may permit an insufficient supply of blood to reach important functional areas, causing an ischemia within this territory, thus producing symptoms less severe than those consequent upon hemorrhage and thrombosis, or with a tendency to show fluctuations in their intensity and duration.

In certain instances, the elastic coat of the artery appears to be duplicated, a condition which in all probability is due to the intramembranous production of connective tissue cells, the proliferation of which delaminates the elastic membrane into several layers. Again, in cases in which the cell proliferation is most active, capillary branches of the vasa vasorum develop from the media and produce new vessels in the thickened intima. In the walls of the thickened arteries the muscle cells of the media ultimately disappear, a process which is one of the steps in the reversion of the highly differentiated arterial structure to the simpler and primitive condition of the tissue from which it springs.

The veins do not entirely escape the changes caused by the infection. There is a uniform thickening with cell infiltration in the vein wall which may eventually determine an obliteration in the lumen.

Syphilitic Meningitis

Gummatous Pachymeningitis.—A meningeal reaction may occur in any or all of the three coverings of the neuraxis. In the involvement of the dura the meningitis is localized and often connected with specific disease of the bone or its periosteum. The pathological change in the dura mater may be of the gummatous variety and characterized by the usual appearance of gumma in other tissues. In conjunction with such gummatous changes there is usually a fibrous hyperplastic meningitis. The dura is thickened to several times its normal size. It is adherent to the arachnoid and pia, from which latter it is inseparable. In many cases blood-vessels are implicated by the pathological process, so that they do not convey sufficient blood supply to their vascular territories. The circulation thus interrupted, causes areas of caseation and necrosis to occur. This condition is *gummatous pachymeningitis.* As a rule, some degree of encephalitis coexists with this change in the dura; and the secondary effects of the infiltration upon the vascular supply may lead to areas of softening in the brain, that is to say, a more or less extensive encephalomalacia, or in the event of involvement of the spinal meninges, a myelomalacia.

Sections of the cortex under these circumstances disclose an infiltration of the perivascular lymph spaces with lymphocytes and plasma cells, extending into the substance of the brain for variable distances. The arteries are usually involved and most frequently manifest an endarteritis, while the neuroglia shows a hyperplasia of varying intensity. Many of the neurones in the vicinity of the infiltration show changes of a disintegrative character, representing various stages of destruction. Medullated nerve fibers also suffer in a similar manner. In lesions of long standing, the nerve tissue is often replaced by a reticulum of fibers and branching neuroglia cells.

Syphilitic Leptomeningitis

The meningeal reatcion may be confined in large part to the pia mater, giving rise to a *leptomeningitis*. This occurs particularly at the base of the brain. In rare instances the arachnoid alone may be involved. The pia and arachnoid are adherent, considerably thickened, and present more or less definite gummatous deposits appearing as solitary infiltrations or as multiple, diffuse infiltrations. These changes are usually associated with an endarteritis, especially when the basal part of the meninges is involved, and the pathological process extends downward into the vertebral canal involving not only the cranial nerves, but also many of the spinal nerve roots. The pia mater is infiltrated by lymphocytes and plasma cells, especially around the vessels and their pial sheaths which extend into the central nervous system. The vessel walls are thickened; a round cell infiltration extends into the perineurium and endoneurium of the cranial and spinal nerves. Microscopically, the distinction between the pia and arachnoid is obliterated by the proliferation of connective tissue.

One of the most important results of the sclerotic changes incident to the basal meningitis is the involvement of the cranial nerves. This pathological change may confine itself to one or two of these nerves, but not a few cases of multiple syphilitic neuritis affecting several of the cranial nerves and spinal nerve roots have been reported.

Localities in which Syphilitic Meningitis is Frequently Observed.—Syphilitic meningitis involves either the base or convexity of the brain. It usually affects one or the other of these areas and is seldom uniformly distributed on the base and convexity. Some mild degree of basal meningitis is found in nearly every case of brain syphilis. The most common site for the syphilitic meningeal reaction is in the interpeduncular space in the region of the optic chiasm. The oculomotor nerves, particularly the oculomotor communis and the optic nerve, are most frequently involved. The process has a marked tendency to extend into the Sylvian fissure and follow the course of the middle cerebral artery. The cranial nerves of the middle fossa of the skull are somewhat more liable to syphilitic involvement than those in the posterior fossa. On the other hand, the syphilitic meningitis may affect the dorsal root of the Gasserian ganglion, also one or both branches of the auditory nerve, together with the facial nerve. If the process extends toward the spinal cord, the ascending portion of the spinal accessory nerve is frequently involved. The nerve roots are affected in various ways. The syphilitic proliferation may make itself most felt upon the nerve fibers, producing *luetic neuritis*. On the other hand, the vascular supply to the nerves may be impeded, due to the endarteritis involving the vessels in the pia mater. The optic nerves are most commonly involved in syphilitic meningitis; next in order of frequency are the nerves controlling the oculomotor apparatus. The facial, auditory and trigeminus nerves are less frequently involved. The vagus, glossopharyngeus and hypoglossus least frequently show meningeal involvement.

Syphiloma (Gumma)

Gummata occurring in the brain, as elsewhere in the body, are large or small single aggregations or multiple discrete neoplasms. In many instances the gummatous process is more diffuse and has the character of an extensive infiltration. Statistical studies of the frequency of gumma involving the brain or spinal cord in cases of late syphilis show a frequency of from 6 to 12 per cent. Fournier in an analysis of 4000 cases of syphilis found gumma in 6.3 per cent. Gron gives a frequency of 11.1 per cent out of 371 cases, and in other statistics shows a frequency of 11.8 per cent in 36,757 cases of lues. The arteries are found to be most frequently the site of gummata, the kidneys and liver next, the brain occupying fourth place.

Syphilomata vary in size from that of a pea to that of a walnut. Occasionally they are much more extensive, large enough, for example, to involve the entire occipital and parietal lobes upon the convexity of the brain. In color they are grayish or reddish gray in the fresh specimen, and sometimes they have a yellowish tint. In consistency they give the impression of rubber-like resistance, from which fact they take their name.

The most common location for the development of gumma in the nervous system is in the dura mater covering the convexity and base of the brain. Upon the basal surface of the cerebrum gummata usually occur as discrete tumor masses, preferably in the neighborhood of the large arterial trunks. The development of syphiloma upon the convexity of the brain seems to give a predisposition to the area of the central convolutions. Frequently the gummata are in direct continuity with the meninges, but in certain cases it is impossible to discover this relation, as the syphiloma may develop in relation with the basal ganglia, internal capsule or deeper structures of the brain. Discrete syphiloma may affect the frontal, parietal, temporal and occipital lobes and may also develop in the fourth ventricle. Among the most common sites for the development of gumma in the brain-stem is the interpeduncular space in a position where the optic chiasm and cerebral peduncles may be simultaneously compromised. The pons, medulla and cerebellum are also sites in which discrete syphiloma has been observed. A number of instances of gumma of the hypophysis cerebri have been reported.

In their surface appearance syphilomata are nodular and irregular. They are usually adherent to the surrounding tissue, particularly the meninges and blood-vessels. In their immediate vicinity there is an attendant local meningitis. Upon section the gumma appears to be a dense homogeneous mass of tissue with a yellowish center surrounded by reddish gray or grayish tissue which may be dense but is often soft in consistency. Upon microscopic examination the characteristic granulation growth of Virchow is observed. Dense accumulations of granulation cells, together with spindle-shaped and star-shaped cells, form the main portion of the gumma. These are imbedded in connective tissue and in places show fatty degeneration. The center of the gumma is usually caseous and necrotic. Its cells are no longer to be distinguished. In places giant cells and large epithelioid elements appear and these may not be distinguished from the giant cells commonly found in tuberculosis. Caseation may be scattered throughout the entire gumma in irregular patches and not confined to the central portion of the mass. The process of fatty degeneration may also be diffuse throughout the neoplasm. It is noteworthy that, according to Nonne, calcification does not occur in gummata.

In many instances the brain tissue itself has the features of a diffuse gummatous infiltration. This infiltration occasions the appearance of a large mass of small round cells with a diffuse distribution which is sometimes referred to as *gummatosis* or *encephalitis gummosa*. The effects upon nerve cells and nerve fibers in cases of gumma and gummatosis are secondary to the compression of the neoplasm in the infiltrating mass.

Syphilitic Meningo-Endarteritis

The pathological changes in meningovascular neurosyphilis are seldom so discrete but that some degree of alteration in the meninges as well as in the blood-vessels may be observed. This possibility justifies the recognition of a syphilitic meningo-endarteritis, a pathological process which partakes of the characteristics both of a syphilitic meningitis and a syphilitic endarteritis. In these cases the dura is generally considerably thickened and more opaque than usual. The Pacchionian bodies show considerable thickening and the pia mater and arachnoid are the site of more or less extensive cellular infiltration. The vessels, especially the larger trunks, show the typical changes of endarteritis—the thickening of the walls, decrease in the size of the lumen, and either complete obliteration of the arterial channel or the formation of a thrombus in the vessel. In consequence, the convolutions over a large area of the brain may be atrophic, a condition which is frequently witnessed in the left inferior frontal convolution in Broca's area. Over this atrophic region the pia is much infiltrated and somewhat edematous.

The meningo-endarteritic variety of syphilis may be characterized by small patchy areas of necroses throughout the brain-stem and the convolutions of the hemispheres due to the partial or complete obliteration of the arterial channels or the development of multiple thrombi. Thrombosis in some of the smaller arteries leads to focal areas of degeneration, a process which seems to have a predilection for the regions supplied by the ganglionic branches of the circle of Willis. The cerebellum and basal ganglia are not infrequently involved by this process.

Syphilitic Meningo-Encephalitis

In addition to the characteristic meningeal infiltration, the brain tissue may show areas closely resembling those of encephalitis. These areas may be scattered through the convolutions of the hemispheres or appear in the brain-stem. Usually they occur as irregular patches not more than a few millimeters in diameter. No gross vascular lesions are demonstrable in connection with these focal areas. The veins, however, in many cases contain masses of polymorphonuclear leucocytes, and the whole region is hyperemic, due largely to the venous congestion. Some atrophy occurs in the medullary substance, probably as the direct result of changes in the cellular substance of the gray matter. The cortical alterations resemble much more closely the pathological changes of an inflammatory reaction than of a diffuse gummatous infiltration, thus justifying the distinction of *syphilitic encephalitis.*

Diffuse Neurosyphilis (Cerebrospinal Syphilis)

This term was devised by Southard under which to classify those cases which pathologically show mesodermogenous changes primarily in the spinal cord and its coverings in combination with syphilitic encephalitis. This variety of neurosyphilis may affect the entire extent of the meninges, both cerebral and spinal, producing destruction in the gray and white matter of the spinal cord and brain. Often the lesions affect the medullary substance of the hemisphere and the corpora striata. With this involvement there is evidence of an acute inflammatory reaction as indicated by a lymphocytosis in the spinal cord, coexisting with meningeal, vascular and parenchymatous changes in the brain.

GENERAL SYMPTOMATOLOGY

Prodromal Period.—The involvement of the nervous system by meningovascular neurosyphilis usually occasions a prodromal period which is more or less characteristic. When the intensity of the lesions centers mostly in the brain, there are changes in the general sense of well-being. The patient's associates notice an alteration in the normal tenor of his life. Irritability, restlessness and insomnia, depression, anxiety and fear, often indicate that some change is in process impairing the individual's efficiency and making clear a definite departure from health. These symptoms during the prodromal period may be variable and fleeting. They may be more or less ingravescent until the first discernible symptom of the disease becomes apparent, and are usually recognizable in the retrospect of the case. Pains in one or another part of the body also have the same variable character during the prodromal period. When the lesion is most intense in and about the spinal cord, the pains are more prominent, especially radiating along the nerves or having a nerve-root distribution. A sense of weakness or failing strength is often noticed, especially in the legs, with easy fatigue and quick exhaustion on exertion. The prodromal period may be so slight as to make but little impression upon the patient, but usually the anamnesis will reveal a stage in the development of the disease when the symptoms in their incipiency are beginning to assume the definite characters which ultimately take form in the well-defined clinical picture of neurosyphilis.

Course and Onset.—The mode of onset in meningovascular neurosyphilis is just as likely to be sudden as gradual. In the cases studied by me, practically 50 per cent had a sudden onset.

In a large number of patients having a gradual onset, the course of the symptoms is steadily progressive with an ingravescence in the severity of the original symptoms and a gradual addition of new somatic or psychic disturbances. A few cases having a gradual onset, manifest a progressive course with occasional remissions. In some instances, characterized by gradual onset, the disturbances come to a standstill and remain stationary or show a slow but steady improvement with some remissions.

Of the cases having a sudden onset, a large number become progressively worse, some slowly and some rapidly, with the addition of new symptoms and an increasing severity of those already present. Such a progressive course is characterized in rare instances, by more or less complete remissions of the prominent symptoms followed by relapses. In a certain percentage of cases having a sudden onset, the somatic disturbances become stationary—show neither progression nor amelioration; while in an equal number the symptoms, which have appeared gradually, improve until the patient attains a state of health approximating the normal. The cases that develop suddenly, occasionally show a tendency to relapses during a slow but gradual process of improvement; while in a small percentage, the cases characterized by a sudden onset show remissions in the symptoms quite as rapid as the onset itself.

In my study of 155 cases, 54 per cent had a progressive course, 23 per cent remained stationary, while 23 per cent showed more or less marked improvement. Remissions during the course of the disease occurred in 9 per cent. Of the cases with remissions, 36 per cent followed a progressive course, 21 per cent were gradually improving, and 43 per cent gave no statement concerning the general course of the disease.

The Initial Symptom

The character of the initial symptom depends largely upon the focus of the most extensive lesion of the disease, whether that be in the nerves, spinal cord, brain-stem or cerebral cortex. It is also influenced by the nature of the pathological process; indeed, the great variability of the initial symptom depends upon whether it is due to increased pressure effects, meningeal irritation, vascular involvement or local infiltration. Any one of the following list of symptoms may appear first in the clinical incipiency of the disease: headache, diffuse or local pains, vertigo, dysesthesia, paralysis and aphasia, tics, spasms or convulsions, defects of smell, taste, sight or hearing, visceral disorders, such as vomiting, palpitation, difficulties of micturition, and psychic disturbances including amnesia, confusion, irritability or mania, depression, hyposomnia, insomnia, hypersomnia, stupor, or coma.

Headache.—The most common of all the initial symptoms developing in the meningovascular type of neurosyphilis is headache. It is likewise one of the most constant symptoms during the course of the disease. It does not occur in all cases, but usually precedes the appearance of other symptoms by weeks, months or years. The head pain is paroxysmal, boring or throbbing. It is subject to periods of remission, and is often worse at night than in the morning. The patient recognizes the headache as a new experience, and even though subject to migrainous attacks, realizes that he is suffering from something more than sick headache.

The attacks may be attended by vertigo of greater or less intensity. The dizziness may be restricted to a mere swimming sensation which is protracted and subject to exacerbations. It is present in some degree most of the time and particularly severe when the headache is at its height. In other instances, during the severity of the headache, vertiginous attacks of great intensity occur, in which the patient is conscious of subjective dislocation of his spatial orientation. He seems to be whirling about from right to left or in the contrary direction; to be suddenly falling to the side backward or forward, while the objects about him appear to pass with great rapidity in the same or opposite direction.

In not a few cases headache is accompanied by vomiting, which is often projectile in character but gives the patient no sense of relief. Prolonged retching may accompany severe headache, lasting for a number of hours. In other instances the initial symptom is headache accompanied by a progressive failure of vision. A severe and prolonged headache which is nearly constant may last for several weeks or months, until the sudden development of one of the major symptoms of the disease, such as hemiplegia or ophthalmoplegia.

Pain.—Frequently the earliest symptom of meningovascular syphilis is pain in other parts of the body than the head. Most often this pain is generalized and may be of a sharp, lancinating character, diffuse in distribution or confined to definite nerve-root zones. The patient complains of a generalized dull aching over the entire body, or of dull pain with a sense of heaviness in both legs. This is especially true in the meningovascular syphilis affecting the spinal cord and its coverings. The pain is usually increased by any exertion and is often worse at night, disturbing sleep and demoralizing the patient to such an extent as to occasion a severe psychoneurosis. The pain may be confined to one shoulder or the other, or extend over one side of the body. Such hemisomatic pain is usually accompanied by dysesthesia and a more or less constant sense of distress or discomfort in the affected side. Frequently the pain is limited to one leg, and seems particularly to affect the course of the sciatic nerve. It is then of a dull, boring or burning character and often so severe as to incapacitate the patient. In some instances the patient complains of pain in the joints. This may be fleeting and transitory, involving all the joints of the extremities, but it more commonly affects the major joints, such as the hip, knee, shoulder and elbow. Pains starting in the shoulder or between the shoulder-blades, and radiating down the arms, are especially frequent as an early symptom in cervical syphilitic meningitis. Cervicalgia or pain in the back and shoulders may occur as the first symptom and persist for a long time before the disease extends and presents symptoms of a more diffuse and general character. Neuralgic pain in the distribution of the fifth nerve on one or both sides, in certain instances, is the first symptom and continues to be the most prominent disturbance due to the disease. It may be of a dull, aching, lancinating or burning character and subject to exacerbations, especially when the face is exposed to cold air; indeed, it may be necessary for the patient to shield the face with some adequate covering, particularly in cold weather. The pain may simulate trigeminal neuralgia and be accompanied by paroxysms in the facial musculature. Usually the patient complains of some degree of dysesthesia in the distribution of the fifth nerve, which takes the form of tingling, itching, numbness or formication. The pain often localizes itself and continues to remain in a discretely circumscribed area, such, for example, as over the eyebrow, in front of the ear or in the anterior nares. It may follow the course of one of the divisions of the trigeminus nerve, or be limited to the territory of a single branch.

Vertigo.— Vertigo in a large number of cases is the first definite symptom of the disease. The dizziness may be unaccompanied by headache or other symptoms and attract the attention of the patient by its frequent recurrence and increasing severity. A sense of instability and uncertainty with swimming in the head may be a constant and distressing symptom. This sensation is often accompanied by nausea and subject to fluctuations in intensity. It is particularly prominent when the patient is fatigued and becomes especially marked on a sudden change of position, as in moving the head quickly from side to side, or attempting to rise from the recumbent to the sitting or standing posture. Acute paroxysms of dizziness also occur, apparently determined by a sudden change in position of the head. These attacks may be followed by intense diaphoresis, sialorrhea, nausea and vomiting. The acute attacks of vertigo may be sudden and shocklike; causing an overthrow of the patient's balance, so that he staggers or falls without warning. Following such attacks the patient is much confused and dazed for a few moments and suffers from a sense of extreme weakness with palpitation of the heart.

Dysesthesia in one or more of the several modalities of sensibility may constitute the initial symptom. These subjective changes in general body sense are usually confined to the extremities and head. They may consist of numbness and tingling in the arms and fingers, a sensation of burning or cold, a sense of constriction about the limbs or body or head, stiffness or a lifeless feeling in the legs, a sensation of numbness of one half of the body and face accompanied by lack of the proper sensory perception of stimuli for touch, pain or vibratory sensibility; a loss of proper appreciation of the position of the legs and less commonly of one or both arms, a sense of constriction about the rectum, this latter symptom occurring particularly with involvement of the lumbrosacral portion of the spinal cord.

Paralysis.—The initial symptom of the disease in many cases manifests itself as a paralysis of the upper motor neurone type. The distribution of such initial paralyses varies considerably. One arm or leg may be involved in a more or less severe monoplegia. In the event of paralysis of the arm there is often an ipsolateral facial palsy of the central type. Often the paralysis is more limited in extent and of less intensity. It may begin as a weakness in the fingers, a weakness in the power of flexion and extension of the hand, a weakness in one or both legs, or a gradual loss of muscular strength in both arms. In a small number of cases it is a loss of muscular power in the shoulder girdle with an atrophy beginning in this region and gradually extending into the arm and forearm.

In certain cases having a sudden onset, the first symptom is a hemiplegia with or without the loss of consciousness, but producing a paralysis of one half of the body. The hemiplegia may develop in consequence of a typical apoplectic seizure; it may on the other hand occur suddenly or after an ingravescent course without the loss of consciousness. Spastic paraplegia frequently appears as the result of spinal involvement, constituting the first symptom in a gradually developing motor disturbance of the upper motor neurone type. The first paralytic symptom may have such a distribution as to involve one side of the body and both legs, that is, a hemiparaplegia, which is most frequent following an apoplectiform seizure. At times the degree of paralysis affecting one side of the body is severe enough only to justify the distinction of hemiparesis. The patient may even give a history of having as his first motor disturbance a tendency to temporary loss of power in the arm or leg, in one side of the body or in both legs, which developed especially when he was fatigued or under nervous strain and seemed to disappear more or less completely after a thorough rest or the absence of psychic excitement. Such transitory paralysis may also affect the cranial nerves, resulting in temporary facial palsies or palsies of the oculomotor mechanism, causing ptosis, transitory diplopia and strabismus.

Sometimes the paralysis representing the first symptom develops in connection with the cranial nerve innervations. The cranial nerve most likely to be affected is the oculomotorius communis, palsies of which result in unilateral or bilateral ptosis, unilateral or bilateral external strabismus with a failure of accommodation and the light reflex, accompanied by a dilatation of the pupil. This ophthalmoplegia may present itself as the first symptom in many combinations with reference to the muscles of the eye involved, but it is most commonly a disturbance of the intrinsic muscles of the eyeball and the levator of the upper eyelid.

Aphasia of the motor type, accompanied by a hemiparesis or, less commonly, a defect in speech of the sensory type producing a jargon aphasia, may constitute the first symptom of the disease.

Spasms and convulsions have occurred in some cases as the first symptom. These spasms may be confined to tonic and tonoclonic contraction in limited areas of the body musculature, such as the arm and forearm or in the thigh and leg. Less commonly they are seen as twitchings and convulsive movements in the facial musculature. Occasionally these paroxysmal attacks have a more extensive distribution and present themselves as typical Jacksonian seizures, having the customary signal symptom, the convulsion beginning consistently in a definite group of muscles and following a constant direction in its spread to the other muscles on the same side of the body. During such Jacksonian attacks consciousness is preserved. In some instances the convulsion becomes generalized and consciousness is lost. Characteristic tono-clonic convulsive seizures, epileptiform in their general nature, appear but rarely as a first symptom of the disease. Momentary loss of consciousness, simulating attacks of petit mal may also be a first symptom.

Involvement of the Special Senses.— The first symptom may declare itself in an involvement of the special senses. A gradual failure of vision or of hearing, or a failure of vision and hearing together, ringing in the ears, color-blindness, tinnitus, vertigo and double vision combined, homonymous hemianopsia, sudden blindness in one eye with pain in the region of the orbit and a paralysis of the internal and external ocular muscles, have all been observed as the initial symptom of meningovascular neurosyphilis.

Visceral Disorders.—Visceral disorders may furnish the first clinical evidence of the disease. Vomiting, with and without headache, may be protracted or recurrent and attended either by profuse diarrhea or constipation. Abdominal pain has also been noted, or a persistent and intractable cough with gastric pain occurring as paroxysms and sometimes followed by hiccough, belching of gas and pain in the stomach, a sense of palpitation in the abdomen attended with palpitation of the heart; dysuria characterized by difficulty in micturition, retention or partial incontinence, pain and burning on urination, urinary urgency with a voidance of a small amount of urine. Impotence, with a loss of sexual desire, is sometimes the first symptom noticed.

Psychic Disorders.—Psychic disturbances are common as the first symptom of the disease. These disorders may result in more or less marked changes in personality, the development of unusual irritability emphasized by irrational outbursts of anger or extravagant expressions of disapproval, a general attitude of irritative discontent, or unusual outbursts of the emotions and irascibility. This state of mind may lead to a marked degree of motor agitation indicative of the patient's discontent. In a few instances such agitation seems to be unconnected with any definite psychomotor incentive and represents a more or less aimless motivation of impulses arising out of indefinite derangement in the sense of well-being. The individual's efficiency and capability may become much impaired by these changes, and the departure from normal may be so pronounced as to attract the attention of his intimates and associates.

On the other hand, the first symptom may present itself as a marked falling off in interest, a lack of attention or power to concentrate, a desire to seek seclusion, an indifference, apathy or even repugnance, accompanied by the attitudes and appearance of depression with all the deprecatory expression of the chronic malcontent.

Hallucinations and delusions, particularly of persecution, inferiority and mysterious influence, may give the first indication of the developing disease. A disturbance of sleep resulting in irregular or protracted insomnia with marked restlessness during the night, may serve as the first clinical signs of a slowly or rapidly developing meningovascular neurosyphilis. Usually with the insomnia and restlessness the patient complains of a feeling of extreme nervousness or of asthenia, of failure of memory and power to concentrate, of general weakness and debility. Occasionally the first symptom is delirium, confusion, mania or stupor. Systematic examination of the patient generally reveals some degree of motor disturbance. These disorders appear in the appendicular musculature but are even more frequent in the muscles innervated by the cranial nerves. Disturbances of somatic sensation and mental status often constitute striking features as a result of the objective physical study of the patient.

DISTURBANCES OF THE NERVOUS SYSTEM ELICITED BY EXAMINATION

Paralysis and Disturbance of Muscle Tone, Volume and Strength.—In a large proportion of cases some degree of paralysis may be discovered on examination. The loss of voluntary motor control may express itself in a slight paresis, most commonly of the upper motor neurone type, evidenced by increased reflexes in the parts affected, the appearance of pathological reflexes, diminution or loss in the superficial reflexes accompanied by a more or less marked increase in the tone of the muscles. The volume and contour of the muscles show little change except such as may be occasioned by disuse of the limb.

The distribution of such upper motor neurone paralysis may confine itself to one limb, commonly the arm. In certain instances, however, the monoplegic defect appears in the leg.

The most frequent form of paralysis is hemiplegia, in which the small muscles of the hand and forearm show the greatest amount of defect in volitional control. The muscles of the foot and leg also give evidence of marked change in voluntary activity, the patient being unable to extend or flex the toes, or to extend or flex the foot. A degree of hypertonus may develop in the affected muscles to produce abnormal attitudes in the upper and lower extremity. The spasticity in the upper extremity holds the hand rigidly in a position flexed at the wrist, the fingers in adduction and flexed at the proximal and distal phalanx, the thumb adducted and held firmly against the index finger or flexed and held beneath the fingers in the palm. The forearm is held flexed at the elbow and the arm slightly abducted from the body, a position especially seen in the patient during locomotion. Abnormal attitudes in the lower extremity are less pronounced, although the foot is often held rigidly in a position of plantar flexion with a slight degree of flexion of the leg upon the thigh. In less extreme cases of paralysis of one side of the body, such abnormal attitudes are wanting, and the loss of strength may be determined only by appropriate tests or observed in the fact that the patient inadvertently uses the paralyzed limb as little as possible.

The hemiplegic defect may essentially disappear only to recur. Such recurrent attacks of hemiplegia sometimes shift from one side of the body to the other. Hemiplegia may develop in the early or in the late stages of neurosyphilis.

The hemiplegia may present itself as a completely ipsolateral paralysis, involving the face, tongue, arm and leg of the same side due to lesions of the cerebral cortex, the corona radiata or the internal capsule. The several varieties of *alternating hemiplegia* also occur, giving rise to the *syndrome of Weber* when the lesion is in the midbrain (paralysis of the eye muscles of one side and the arm and leg of the opposite side), to the *syndrome of Millard-Gubler* when the lesion is in the pons (paralysis of one side of the face and opposite arm and leg), or to the *syndrome of lingual alternating hemiplegia,* when the lesion is in the medulla (paralysis of one half of the tongue with paralysis of the opposite leg and arm). Other more irregular forms of hemiplegia also occur in combination with one or more of the cranial nerves.

Budd cites 3 cases of hemiplegia which resulted from apoplectic seizures occurring in the different stages of constitutional syphilis.

When the lesion confines itself to the spinal cord in its most usual site of involvement, namely, in the lower thoracic segments, the paralysis is paraplegic in its distribution, involving both legs. In this instance there is a marked increase in the tonicity of the muscles, together with an impairment in volitional control. Certain cases, usually of the congenital type, show some degree of paralysis of the upper motor neurone type in all four extremities. The lesion upon which this disturbance is based is an agenesis or disturbance in development. Under such conditions the muscles in the upper extremity at first show hypertonicity with abnormal attitudes and increased reflexes. Later these muscles begin to atrophy, and contractual deformities appear in the arms and hands. At this time spastic paraplegia usually develops. Examination of the patient may reveal the fact that a spastic paralysis has involved both legs and one arm, thus producing a loss of power having a hemiparaplegic distribution.

In contradistinction to the upper motor neurone type of paralysis which is characteristic of the great majority of patients suffering from meningovascular neurosyphilis, a flaccid paralysis occurs in some instances. This paralysis is accompanied by a decrease or disappearance of the deep reflexes, a loss in the contour and volume of the muscles in the affected parts, and a marked reduction in the muscular tone. Paralysis of this kind occurs in the leg in consequence of a syphilitic neuritis of the sciatic nerve. It may also affect one or both arms, either as the result of a syphilitic neuritis or a syphilitic radiculitis. Less commonly, syphilitic endarteritis involves the ventral gray columns of the cord in the cervical or lumbrosacral region, and produces a flaccid paralysis with loss of reflexes, resembling in all its principal aspects the motor disturbance of anterior poliomyelitis. Paralysis of this kind is usually slowly progressive and tends to extend from one segment of the limb to another and gradually to involve large areas of the body musculature.

Gait.—In a certain percentage of cases examination of the gait shows a disturbance in locomotion. In my series of cases 23 per cent showed defects in gait. These disturbances may be slight, arising from weakness in the muscles of one or both legs; but often result from such a marked degree of spasticity as to render walking barely possible. In lesions affecting the spinal cord, the gait usually presents a paraplegic defect with a tendency to cross-legged progression. The rigidity in the legs also produces a columnar effect in walking, each leg being moved forward stiffly, the foot dragged along the ground with a shuffling sound, the toe turned inward and oftentimes with a loss of the normal heel-toe time element, the heel striking the ground after the toes or simultaneously with them. In rarer instances of disturbed locomotion the heel does not come to the ground at all, the degree of spasticity being such that each step produces several oscillations up and down, the toes being turned inward and the knees rubbing against each other or one leg crossed in front of the other. The defects of the spastic paraplegic gait are often augmented by a marked incoordination, causing the patient to stagger, and producing an ataxic spastic paraplegia.

Lesions producing paralysis of one side of the body give rise to a more or less pronounced hemiplegic gait, with a dragging of the paralyzed leg, the toes turned out in eversion and dragged along the ground with an audible shuffle at each step. In addition to this disorder in the lower extremities, the automatic associated movements normally observed in the arm are greatly altered. The paralyzed arm, with each step of the sound leg is held in abduction from the body, the forearm flexed at the elbow with a considerable exaggeration of the backward and forward movements of the arm.

Occasionally the gait is disturbed by a peripheral paralysis often affecting the anterior tibial nerve and giving rise to steppage gait with foot-drop. The lesion may involve the ventral column cells of the spinal cord, producing a bilateral lower motor neurone paralysis with double toe-drop, and in a few cases it affects the innervation of the gluteal and hamstring muscles in such a way as to produce a waddling gait in many respects similar to that seen in dystrophies.

Coordination.—Equilibratory coordination is often disturbed in this form of syphilis, the patient showing a Romberg sign in about 16 per cent of cases. This disturbance appears to be due to interference with the sensory tracts from the proprioceptive system, largely those concerned in muscle-joint sensibility. The patient finds difficulty in standing with the feet together and eyes open; this act is even more difficult with the eyes closed. Standing on one foot or the other also develops some degree of disturbance in maintaining balance. Coordination not concerned in equilibration suffers in a small number of cases. The finger-to-nose, finger-to-finger tests, particularly with the eyes closed, as well as the pointing tests, all develop slight inaccuracies in the synergic control of the muscles. These defects are most prominent near the end of the movement, as the patient is about to reach the goal of the act. The heel-to-knee tests also develop some degree of inaccuracy in the synergic action of the muscles. Adiadochokinesis (inability to perform succession movements with equal facility in both upper extremities or in the feet and legs) has been observed, but is of rare occurrence. Dysmetria is equally infrequent, although some slight degree of disturbance in the measurement of distances of skilled acts is observed in many cases. It should, however, be considered as a mild degree of asynergia.

Reflexes are variously affected. In 65 of the cases considered in this series, the deep tendon reflexes were recorded as being normal; in 44 the responses were increased, while in 8 there was a general decrease in the activity of the response. In 11 cases the knee-jerks were recorded as being absent, while the other deep reflexes were either normal or somewhat depressed. The absence of the knee-jerk, unassociated with muscular atrophy or fibrillation, is an evidence of involvement of the dorsal root by the meningovascular process. In other cases, when there has been atrophy and fibrillation of the anterior thigh muscles, the loss of the knee-jerk may be attributed to the morbid process attacking the ventral gray matter of the cord. In the great majority of cases, however, the loss of the knee-jerk is dependent upon the interruption of the sensory side of the reflex arc— a process of strangulation of the dorsal root as it passes through the membranes and approaches the cord.

In 26 of the cases the reflexes were unequal, usually a definite hyperexcitability of one side as compared with the other. In many of these cases this phenomenon was associated with a loss of the superficial cutaneous reflexes of the abdominal wall and the presence of pathological reflexes on the same side. This combination indicates the presence of some sort of hemiplegic accident in the course of the disease, either spinal or cerebral in location.

The *superficial reflexes* were absent in 11 and unequal in 13 of the cases recorded. This loss or inequality was almost constantly associated with other evidences of hemiplegic defects. In a certain number of cases the loss of the superficial abdominal reflexes seemed to be a part not of a hemiplegic defect but of a diffusely scattered multiple lesion symptom-complex.

Abnormal reflexes were present in 23 cases. Usually the Babinski reaction alone was present, but in some instances all the corroborative tests were present also. Usually, as mentioned above, the Babinski reaction was associated with other evidences of hemiplegic defect, but in certain cases it evidenced a widespread involvement of the spinal cord, being bilateral in 4 cases.

Skilled acts are frequently disturbed. Although apraxia in the strict sense is not a common symptom, some degree of disorganization in the control of skilled movements is observed in many cases. This more especially affects the skilled acts necessary to speech, producing dysphonia or, in more extreme bulbar cases, aphonia. When the lesion in the speech areas is cortical or subcortical, aphasia in one of its several forms occurs. In my series of cases 13 per cent presented a sensory or motor aphasia. The speech disturbance may amount only to a slight disorder of the sensory type in which spontaneous speech is disturbed, together with the power to enumerate and denominate properly, an inability to speak from dictation or read aloud, to read from dictation, the misuse of words, speaking ungrammatically out of proportion to the patient's education, perseveration, or the use of an actual jargon. In other instances the speech defect is more distinctly motor, the sensory control and understanding of spoken language being retained with more or less complete power of emissary speech. The impairment is usually accompanied by some degree of defect in handwriting.

Abnormal Involuntary Movements.—The most common form of abnormal involuntary movement occurs as localized spasms or more extensive movements involving one half of the body in the manner of a Jacksonian attack, or as a generalized convulsive seizure. Isolated spasms in the arm or leg occur most frequently as the result of meningeal irritation to the cortex. This also is the case in the hemisomatic convulsion involving one side of the body. Vascular lesions involving the deeper structures of the brain also give rise to this type of seizure. Generalized convulsions are of common occurrence in meningovascular neurosyphilis as well as in parenchymatous forms. This disturbance may be the result of a diffuse syphilitic endarteritis involving the deeper structures of the brain, or a diffuse syphilitic meningitis of the convexity and base of the brain. Tremors of a fine and rapid variety in the extremities, tongue and face appear occasionally in patients suffering from diffuse syphilitic endarteritis and are most pronounced during exacerbations in the course of the disease. In a few instances a tremor resembling that of paralysis agitans in rate, rhythm, amplitude and distribution, has been reported in connection with neurosyphilis. Myoclonic and fibrillary twitchings in the muscles, although rare, appear in connection with the spinal form of the disease, due to involvement of the ventral gray columns of the cord or the nerve roots. They are indicative of a degenerative process compromising the integrity of the muscular tissues. Athetoid and choreiform movements, although rare, have been observed as the result of neurosyphilis. These disorders are now believed to be due to involvement of the corpus striatum, the red nucleus or subthalamic region of the interbrain.

Abnormal associated movements are of frequent occurrence in neurosyphilis, especially in upper motor neurone paralysis of the hemiplegic type. Observations concerning these pathological associations in motor activity are of such interest in all cases of hemiplegia as to warrant more extensive attention than they usually receive. The paralyzed side of most syphilitic hemiplegics will manifest upon examination a number of these abnormal associated movements. Among these are the ***pronation sign of Strümpell,*** which consists of pronation of the forearm on passive flexion at the elbow; the ***thumb sign of Hoffmann,*** consisting of flexion of the thumb on snapping the finger nails of the paralyzed hand; the ***thumb sign of Klippel-Weil,*** consisting of flexion of the thumb on passive extension of the fingers; the ***trunk-thigh sign of Babinski,*** consisting of elevation of the affected leg determined by the effort of the patient to sit up from the recumbent position, the ***reinforcement sign of Babinski,*** consisting of elevation of the affected leg when pulling on the hand; the ***tibialis sign of*** *Strümpell,* consisting of the dorsal extension of the foot on flexion of the leg at the knee; the *coughing sign of Huntington*—the sharp elevation of the affected leg when the patient coughs. All of these signs are to be regarded as confirmatory of an upper motor neurone lesion, usually in the intracerebral portion of the pyramidal course.

Disturbances in Sensation.—Sensory disturbances, though less common than motor disorders, were found in 18 per cent of my series of 155 cases, and this proportion is in general accord with that of other observers. As a rule, the most marked sensory disturbances are those occasioned by spinal cord and nerve involvement. Subjectively, they consist of dysesthesias and paresthesias, that is, abnormal sensations of which the patient complains, such as numbness, tingling or burning, or the perversion of sensory stimuli in such a manner that a pinpoint produces a sensation of burning, warmth or cold a sensation of pain, touch and light pressure a sensation of pain, and deep pressure a sensation of burning. Occasionally the distribution of the sensory disturbance is hemisomatic, in consequence of a cortical or subcortical lesion in the parietal area of the brain. In such instances all qualities and modalities of sensibility are affected. In lesions limited to the higher associative areas of body sensibility (superior parietal convolution) hemi-astereognosis occurs, as the result of which the patient is unable to identify by palpation the form, size, weight and consistency of familiar objects. Deeper lesions situated in or near the posterior limb of the internal capsule produce a similar astereognosis in combination with homonymous hemianopsia due to involvement of the optic radiation. Rare cases in which the major pathological change is confined to the lateral part of the thalamus show marked sensory changes characteristic of the *thalamic syndrome* in which there is a loss of acuity in all types of sensibility, the presence of spasmodic pains in one half of the body, and a peculiar elevation in the affective reaction to stimulation of the anesthetic areas of the body. Pain in association with diminution of sensory acuity or even anesthesia may occur, having a peripheral distribution in nerve zones or root and segment zones, as in radicular and spinal involvement. This latter change is indicative of morbid processes in or about the dorsal root at its point of entrance into the spinal cord.

In only a small proportion of the cases was there any evidence of disturbances in the deep sensibilities. Vibratory sense was impaired or lost in about 2 per cent of the cases recorded. The sense of movement, muscle-joint and tendon sense were involved in only a few cases, and in these cases affected more the lower than the upper extremities. In general, the deep sensibilities were relatively infrequently affected, and when affected the disturbance was comparatively slight.

Cranial Nerves.—One or more of the cranial nerves are frequently affected in neurosyphilis, especially in the diffuse and cerebral forms. In my series of 155 cases, involvement of one or more of the cranial nerves occurred in 70 per cent. Of these the optic, oculomotor and facial nerves were most often involved.

The *olfactory nerve* on one or both sides showed departure from the normal in 2 per cent of the cases. This involvement resulted symptomatically in a reduction or loss of the sense of smell. In addition to this anosmia, cases of perverted olfactory sensibility are observed, and also olfactory hallucinosis. When the latter is present the patient complains either of disagreeable or agreeable odors, more or less constant, without existence of the corresponding stimulus in the environment. This sensory hallucination is subject to exacerbations which frequently terminate in a peculiar state of mind, giving the patient a sense of unreality or remoteness, and creating an indefinable impression in his mind that he has experienced similar circumstances frequently in the past. This sense of remoteness and reminiscence culminates in a feeling of distress and may be attended by a generalized convulsion. The entire symptom-complex, beginning with the olfactory hallucination and terminating with the convulsion, is known as an *uncinate fit.* It is due to a lesion in or near the uncus situated at the tip of the temporal lobe.

Optic Nerve.—Neurosyphilis of the meningovascular type is often acompanied by symptoms referable to pathological changes in the second nerve. The clinical signs of this change are manifest as papilledema, optic neuritis, neuritic atrophy and simple atrophy. In the majority of cases these changes in the optic nerve are associated with basal meningitis. Choked disc, when present, usually affects both eyes, although the papilledema may be greater in one disc than the other. Optic neuritis, on the other hand, frequently remains unilateral. Choked disc is generally accompanied by marked increase of intracranial pressure, while the neuritis is indicative of an extension of the syphilitic process into the optic sheath and finally into the optic nerve itself. Gummatous meningitis may primarily affect the optic chiasm or one of the optic tracts, in consequence of which visual disturbances exist without demonstrable changes in the fundus.

Hemianopsia, either bitemporal in its distribution and due to involvement of the optic chiasm, or homonymous, due to a lesion in one of the optic tracts or optic radiation, is not infrequently seen as a result of syphilitic endarteritis, basal meningitis or thrombosis. Homonymous hemianopsia also occurs from large gummatous areas or areas of softening involving the occipital lobe. In rare instances the distribution of the defect in the visual fields is that of a binasal hemianopsia.

Syphilitic endarteritis, gummatous growths and diffuse meningitis extending along the course of the optic fibers and compromising vision are a common occurrence. Syphilitic involvement of the peri-orbital fascia of one or both eyes may produce sudden blindness preceded by severe headache and followed by optic atrophy. A change in the optic nerve secondary to neuritis or perineuritis, caused by a gummatous process in the base of the brain, may produce a *neuritis descendens*. This frequently affects the macular bundle in the nerve and produces a central scotoma with blindness or blurring in the center of the visual field. In certain cases the deeper fibers of the optic nerve are involved, the macular bundle escaping pathological change. As a result, diffuse or peripheral scotoma is produced, with all grades of partial blindness. It is obvious from these facts that all degrees of amaurosis occur in consequence of a syphilitic involvement of the optic mechanism, varying from limited scotomata in the center or periphery of the visual fields to complete blindness.

The pathological changes affecting the receptor system of the optic mechanism, particularly the retina, may result in syphilitic retinitis which usually develops in the secondary stage and exists apart from a chorioiditis. The macula is most frequently the seat of the involvement, although any part of the retina may be affected. Syphilitic retinitis is also seen in congenital syphilis. *Retinitis proliferans* is a rarer pathological change in patients suffering from neurosyphilis. It sometimes follows syphilitic retinitis. The vision is greatly impaired and may be entirely lost. The development of new tissue by its contraction causes puckering and detachment of the retina.

Chorioiditis is of more frequent occurrence than retinitis. Although the close association of the choroid and retina is conducive to involvement of the latter in chorioiditis, the retinitis is usually secondary.

Papillitis, with some degree of swelling of the nerve head, is often seen due to gummata developing in the basal portion of the brain. Increased intracranial pressure is undoubtedly responsible for the disc changes, although a meningitis of syphilitic origin is capable of producing these disturbances.

Thirteen per cent of my cases showed pathological changes in the fundi; 33 per cent were noted as normal, while in 54 per cent no statement as to the condition of the eyegrounds was made.

Hughlings Jackson describes a case of atrophy of the optic nerve occurring in a boy, due to congenital syphilis, and another case of necrosis of the frontal bone and atrophy of the optic nerve in acquired syphilis. The sudden loss of sight was unaccompanied by evidence of local disease in the eye such as is generally recognized as syphilitic.

[*To be concluded in the November number*]

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BRADYKINETIC ANALYSIS OF SOMATIC MOTOR DISTURBANCES

ANALYSIS OF MOTOR DISORDERS BY THE AID OF ULTRA-RAPID MOVING PICTURES

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The subject of disturbed motor function has been carefully studied for many years with some interesting results especially in the clinical field. Motor defects, however, have proved elusive to exact interpretation and in many respects knowledge in this particular field is still in need of assiduous cultivation. It seems probable that the principal difficulty lies in the present imperfect understanding of vertebrate motion as a whole, and in this light the interpretation of disorders in the complex motor mechanisms of man appears all the more perplexing. It has been apparent to the authors and others interested in this important topic that only by a combination of many different methods may the problems involved in the motor disturbances be solved. The object of this paper, therefore, is to present certain newer methods of study which may be applied to these disorders, and at the same time to demonstrate the results derived by an application of the methods to certain well-defined groups of motor disorganization.

Weisenburg, a few years ago, demonstrated the value of cinematographic reproduction in the study of nervous diseases. In the present investigation more recent developments in photography with the high speed-camera are introduced in the analysis of deformities in motion.

In an attempt to analyze and record the disturbances of motility, one is impressed at the outset by the inadequacy of the nomenclature employed in describing various types of abnormal motion. Most of the terms in common use not only fail to be descriptive, but lexicologically many of them are nondescripts. For the present and until adequate replacements in terminology may be made, it seems desirable to adhere to the accepted nomenclature.

METHOD AND MATERIAL

Ultra-rapid moving pictures of patients by means of the "Novograph" camera were taken at the rate either of one hundred and sixty or three hundred exposures per second. The ordinary cinematographic rate is sixteen exposures per second, so that the ultra-rapid rate is from ten to eighteen times as fast. The normal rate of projection on the screen offers opportunity for the study of every component in each movement. The motion, thus slowed down, serves as the basis for what we have called *bradykinetic analysis.*

In studying each of these types photographs were made simultaneously at the usual and also at the ultra-rapid rate of exposure. The screen reproduction of these pictures first shows the subjects photographed at the regular cinematographic rate, followed by the pictures taken at the ultra-rapid rate, both sets of photographs being reproduced at the usual cinematographic rate of sixteen pictures per second.

The material upon which these studies were made consisted of patients selected from the neurological wards of the Montefiore Hospital for Chronic Diseases. In this selection special attention was given to securing patients showing marked motor disorders the general characters of which have been the subject of much debate and have also occasioned not a little discussion in classification and nomenclature. The motor disorders submitted to bradykinetic analysis include choreiform movements, torsion spasms, extreme intention tremors and cerebellar ataxia. These motor defects occurred in cases of *chronic degenerative chorea, astasia abasia, dystonia musculorum deformans, multiple sclerosis* and *congenital cerebellar aplasia.*

In all the cases studied the motor defect was characterized by some pronounced abnormal and adventitious element of motion which produced a definite deformity of movement. Such adventitious motor elements we have endeavored to describe as objectively as possible in the following enumeration of them:

1. Regular and rhythmical oscillations of parts of the body during muscular inactivity or in volitional action.
2. Irregular arrhythmical oscillations of parts of the body during volitional motion.
3. Sudden isolated twitches of the muscles producing involuntary movements or deflecting the course of volitional motion.
4. Sudden successive twitches of muscles producing involuntary movements or deflecting the course of volitional motion.
5. Sustained mobile spasms of muscles during volitional motion producing rhythmical distortions of the limb, trunk or limited parts of the body during activity of the particular part.
6. Sustained tonic spasms of limited groups of muscles during volitional motion which retard or deform the resulting movements.
7. Sustained tonic spasm in a large number of muscle groups during activity or in rest, producing distortion of the body together with marked deformity of motion during action.

I. ANALYSIS OF A CASE OF CHRONIC DEGENERATIVE CHOREA

This patient was a married woman, thirty-six years of age, born in Russia. She was the mother of five healthy children. Her present illness began when she was thirty-one years old during the fifth month of a pregnancy. Without apparent cause there developed irregular, purposeless, involuntary movements in all segments of the body. A gradual change in conduct was also observed which apparently affected the faculties of attention and concentration. She showed lack of interest in her family and in affairs generally, with a progressive mental deterioration.

The cinematographic study of the patient at the normal rate of projection showed a variety of meaningless and purposeless deformities of motion such as facial grimacing, continuous movement of the eyeballs together with sudden twitchings in the trunk, limbs and head. These movements were somewhat too rapid for the typical Huntington's chorea. There was incessant jaw motion as in grinding the teeth, and the tongue and lips smacked at irregular intervals. The abdominal muscles were in almost constant play.

By ultra-rapid photography, the projection revealed striking differences for interpretation. The component elements of the deformities of motion were clearly discernible, and what appeared to be twitching, purposeless movements were seen to have definite associative significance. They comprised a series of symbolic attitudes indicative of emotion. The photographic analysis reduced the speed of movement and resolved the motions of the trunk and extremities into segmental components. These movements now appeared as those of approach and retreat, expressing the appeal of entreaty or negative self-feeling of shame. They clearly represented an emotional flux recognized as a functional element in some patients with this type of chorea.

The posing and attitudinizing, so impressive in other well-recognized organic diseases, suggest a close anatomical relation of these symbolic movements to the basal ganglia of the forebrain. The thalamus and the corpus striatum which are functionally associated with sensorimotor complexes serve as the centers especially controlling automatic associated acts and attitudes expressing the fundamental emotions. In this light it seems fair to presume that such attitudes as express emotional states in true hysteria may have their subconscious controlling level in these basal ganglia.

Another interesting feature in the bradykinetic analysis was the extreme hyperextension of the left leg at the knee as the patient walked. In this patient the attitude of the limb appeared to be the result of kinetic overflow within the extensors, the check of the antagonistic flexors being defective. The extreme slowness of the antagonistic check response together with a singular wave reaction seen in the extensors suggested a possible failure of elements constituting the so-called static mechanism. A similar failure in stabilizing the kinetic flow is seen in the study of the dystonias, and the imbalance in the synergic units points to a defect in the synergizing influence of the cerebellar reflex circuit.

In summary, the analysis of this case of degenerative chorea discloses a combined defect, in part affecting the kinetic mechanism controlling automatic associated movements and attitudes expressive of the emotions, in part due to impairment in the static mechanism maintaining posture. (Fig 1.)

II. ANALYSIS OF A CASE OF CHRONIC DEGENERATIVE CHOREA WITH SOME ELEMENTS SUGGESTIVE OF DYSTONIA

The patient was thirty-six years of age. About two years previous he became "extremely nervous" and developed slight twitching in the lower extremities, soon accompanied by a staggering and uncertain gait. Four months later the upper limbs participated in motor restlessness, consisting of involuntary "flinging" about of the arms followed by shrugging movements of the shoulders. The same disturbance involved muscles of the head and neck. A corkscrew-like motion, chiefly of the trunk, completed the motor disorder.

When admitted to the Montefiore Hospital he had the attitude, movements and gait of Huntington's chorea.

By bradykinetic analysis the attitudinizing and symbolization, so evident in the first case, were apparently distinctly those of hurling or lifting and were too definite in pose and in poise to be a mere chance overflow of kinetic energy into purely accidental designs. Unquestionably there was an element of purpose in these attitudes. The postures were those of aggression and attack, thus expressing what may be expected of the more positive character of the male emotive mechanism. An extensor elevation of the foot (not seen in the accompanying photographic illustrations) as the patient stepped forward, and also a corkscrew-like movement of the trunk upon the pelvis, appeared to be distinctly dystonic elements and suggested a relationship between the dystonias and this type of chorea.

In all the cases presenting clinical features that seem to involve the posturing and attitudinizing mechanism, the motor elements which determine posture seem to be disorganized. In cases of this type the agonist and antagonist are apparently not harmoniously cooperative. This becomes manifest as the several muscles are studied in their slow interplay. Comparison by means of bradykinetic analysis readily demonstrates a marked difference between disturbances due strictly to cerebellar disintegration of the synergic unit, and those forms of striatal disintegration whose pathological anatomy probably lies in the corpus striatum and its immediate basal connections, especially with the red nucleus.

As many observers have pointed out, there is doubtless an intimate anatomical pathological relationship between the various deformities of motion of the striatal type, and in all of our cases a study of the muscle-group activities brings clinical confirmation of this fact.

The prolonged pronation spasms in this patient and the marked overaction of the extensor muscles holding the raised foot upon the toes, clearly belonged to the group of sustained mobile spasms producing rhythmical distortions of the limbs and body during motion. This phenomenon is particularly prominent in the dystonias.

The tendency of trunk distortion was disclosed as a sudden unilateral pull of flexor trunk groups. The torsion in this case involved rather the upper trunk segment in contrast to that seen in dystonia. There was a suggestion, too, of tic element in the choreic movements which involved especially the axio-appendicular groups of the upper extremities. Analysis certainly demonstrates a variety of elements common to many entities. (Figs. 2 and 3.)

In summary, the analysis of this case of degenerative chorea discloses the fact that sudden isolated twitches of the muscles producing involuntary movements or deflecting the course of volitional motion are to be associated with disturbance in the striatal and cerebellar mechanisms, and that the attitudinizing, although different from that observed in the woman suffering from the same disease, is basically similar in being expressive of primitive emotions. In the case of the male these emotions have a positive self-feeling tone representative of aggression and attack in contrast to the female where these emotions have their expression as negative self-feeling tone in reactions of appeal and retreat. The sustained tonic spasms producing distortions are manifestations of hyperkinetic innervation of muscles recognized as the essential part of the syndrome in those motor disorders known as the dystonias.

III. ANALYSIS OF A CASE OF MULTIPLE SCLEROSIS WITH ASYNERGIC MOVEMENTS

The patient, a watchmaker, forty-eight years of age, first noticed, when he was thirty-two years old, a slight and occasional defect in vision, with numbness in his fingers which occurred from time to time. A year later he began to be troubled with a tremor in his right hand especially when executing the more delicate manipulations of his trade. This tremor gradually spread until it involved his entire body. He soon developed difficulty in walking, and in the course of three years his locomotion was so much compromised by a spastic paraplegia that he was forced to give up his occupation. It was about this time that the nature of his malady was recognized.

The study of this case at normal photographic speed discloses many confusing movements as the subject attempts the "finger-to-finger" approximation test. He illustrates particularly well a phenomenon which, in part, characterizes the motor disturbances of disseminated sclerosis, the so-called *intention tremor.* These abnormal movements during voluntary effort were, from an early period in the history of his disease, very extensive, and comprised, indeed, the only symptom present for some time. Original observers of the case placed it among functional disorders.

The analytic projection shows a disintegration of the synergic units throughout the body. Agonist and antagonist are not coordinated. The extensor check fails to hold the flexors and the reverse is also true.

This disorganization is especially obvious in the finger-to-finger test as the point of destination and finer approximation is approached. In slow motion the disproportion in flexion, extension and supination is clearly seen. The arms shoot far beyond the point desired (the analytic photography greatly reducing the speed), they are overextended and again overflexed. The obtrusive feature is a motor disorganization showing dissociation of muscular harmony and asynergia between dominant and check groups of muscles. As has been shown by Weisenburg and Tilney, this type of motor phenomenon is indicative of cerebellar disturbance. The groups of muscles of the trunk and extremities are also involved. The case is one of integrative asynergia. It would seem then that the "tremor" of multiple sclerosis is of cerebellar origin, that a case such as the one studied must involve extensive cerebellar areas. (Fig. 4.) An even more definite conception of cerebellar asynergia is acquired by a study of a case of cerebellar agenesis.

In summary, the analysis of this case of multiple sclerosis reveals the fact that the so-called intention tremor is a static defect due to the dissociation of the synergic units of the body consequent upon disproportionate innervation of the dominant and check elements in the muscle groups.

IV. ANALYSIS OF A CASE OF ASTASIA ABASIA WITH EXCESSIVE ISOLATED AND SUCCESSIVE SPASMS

The patient was a young girl, seventeen years old, apparently with nothing in her family history to cause the onset of a profound psychoneurosis. The abnormal movements consisting of isolated and successive muscular spasms had their exciting cause some four years previous in the fear of a thunder storm. Associated with these movements was a psychic attitude of dread with sudden increase of the motor agitation and facial expression of anxiety upon any sudden noise. Up to that time there had been no disturbance of consciousness nor had there been objective or subjective sensory symptoms of hysterical nature.

The motor manifestations were those of astasia abasia, the patient being unable to stand or walk without support.

At normal speed photography, as the patient stood and walked with support, the gait was not suggestive of any organic nervous disease. The segments of the body, as the muscles moved the joints, acted in harmony and unison; there was no disintegration of the synergic units. The lower extremities in walking performed a series of short steppage movements, and in standing these continued in rapid succession. Analyzed under increased photographic speed, rhythmic, well poised, fully coordinated, normally synergic phases of voluntary movement followed in regular order. Their contrast with the preceding studies of organic deformities of motion leaves no doubt of the different character of each. These movements clearly bespeak an intense self-consciousness with acute apprehension of impending danger. They are more complex and studied than the more direct instinctive reactions to fear. Their elaborate character is in marked contrast to the simpler attitudinizing in chorea.

Another muscular phenomenon was observed in this case, namely, a vermicular muscle wave. This was not the result of any artificial or mechanical effect and must have depended upon some inherent quality of the muscle groups. The muscle bellies themselves were apparently the seat of gross morphological changes. Whether this phenomenon is pathognomonic of functional motor disturbances can be determined only upon bradykinetic studies in many other cases. Mr. C. P. Watson, President of the Novograph Company, whose experience with screen photography is vast, and who had witnessed film productions of many athletes and others under varying conditions of muscular activity, had not previously observed this vermicular wave. In the normal projection of this girl there was no suggestion of this wave, but in slow motion analysis the wave was immediately recognized passing slowly from below upward through all the muscles of the right lower extremity from ankle to hip. This appearance resembles the successive surface movements when a strong wind sweeps a field of wheat. It would be of interest to study by bradykinetic analysis a number of functional disturbances in which the somatic musculature is involved. Possibly this wave as demonstrated in this patient is a condition found only in what we are wont to call non-organic or functional disorders.

Note: Some few weeks after the photographic presentation and following a fright, the patient suddenly passed into a kind of hypnotic sleep from which she could not be aroused even by the strongest stimuli, including faradic current of great intensity. In the beginning of this state the girl screamed continuously, had tonic and choreic convulsive seizures and assumed bizarre, emotional attitudes. She would constantly repeat, "I am afraid," but could not be aroused to any form of conscious response. There was corneal as well as general anesthesia. The condition continued for some three or four days. She was regularly tube-fed and catheterized. The irregular convulsive seizures were really a marked intensification of previous movements with an intense psychic phase added; they were so violent as to yield only to chloroform narcosis pushed beyond the primary stage. The patient at no time showed signs of exhaustion. There were several periods of these seizures, each lasting a few days. It seems to be of some significance that the patient was restored to consciousness by the playing of phonograph records. A more careful study of the intricate psychic associations was not made. The patient has since markedly improved psychically, but it is of interest to note that the toes in which intense flexor movements were observed on the screen have since assumed a definite and fixed attitude in the flexor position, though in sleep they at times become relaxed. (Fig. 5.)

In summary, the analysis of this case of astasia abasia reveals the fact that the irregular, sudden twitchings of the muscles which disturbed locomotion were in reality highly complex and expressive of acutely conscious and psychically much magnified fear. These movements are in marked contrast to the simpler attitudinizing of chorea. The wavelike contraction in the muscles of the leg requires further study before a complete analysis of it may be offered.

V. ANALYSIS OF THREE FAMILIAL CASES OF DYSTONIA MUSCULORUM DEFORMANS (A BROTHER AND TWO SISTERS)

We are indebted to Dr. I. Abrahamson for the opportunity of studying this series of cases.

In this family group of dystonia musculorum deformans, each member presented a different degree of severity and progress of the disease. The family stock was recorded as having been substantially without taint except in one important particular: On the paternal side there was a more or less direct history of dystonia, in that the patients' father's sister and her daughter were afflicted with a disease similar to that of the three patients here described.

1. *The Brother. Incipient Stage. Movements Largely Confined to One Extremity.* The boy was the least afflicted of the group. As in the case of the sisters, his early life up to the time of onset of the present illness was quite normal. The motor disturbance began in his thirteenth year, and as in the case of the younger sister, manifested itself especially in an attempt at execution of finer movements with the right hand, such as writing. The patient was intelligent and had made from the first every effort to overcome his difficulty. He fully realized the progress and serious deformity which the disease had shown in his sisters and was fearful, even before the onset of his illness, that he too might become a sufferer. Indeed, the boy made unusual efforts by exercise and intense mental endeavor to maintain control over the afflicted upper extremities; this he continued with great determination as he felt the right leg, more particularly the right foot, assume spasmodic, uncontrollable, involuntary attitudes in attempting movement.

The cinematographic representation of his gait showed a turning-out of the foot and the sudden flexion at the knee and hip with inward rotation at the hip. In normal projection as he ran, a limp alone was seen. The slow motion for bradykinetic analysis showed a partial failure of coordinate time reaction in groups of agonists and antagonists. A study of our series of dystonias shows this failure as a feature in them all. For example, the forceful hyperextension of the foot, common to all of the cases of dystonia, appears as a failure in the time limitation of the muscular contraction, or, in other words, an excessive overflow of innervation. The peculiar character of this kinetic overflow in the agonist groups differentiates the organic from the functional disease with which this latter spasmodic condition has often been confused. From the standpoint of muscular dynamics, this excessive muscular contraction is wasteful and unnecessary for the purposes served by the muscle action. A small percentage of energy is expended in the purpose and a large amount in producing useless tonic spasms of the muscles involved.

The slight rotation of the thigh seen on normal projection, together with a slight raising of the foot in equinus position and flexion at the knee and hip, was in reality a widespread disorganization of the movements of the gait. Analysis showed at once that this form of deformity was due to excessive overflow of kinetic innervation.

Any movement designed to serve some definite purpose, as for example, moving the leg forward in walking, may be considered as a transition from an *initial posture to terminal posture*. The leg may be arrested at any intermediate point and be made to assume an *intermediate posture*. The motion, therefore, from the initial to the terminal posture may be regarded as a series of potential intermediate postures between the two limits mentioned (initial and terminal posture). The innervation producing motion through a series of postures from the initial to the terminal is *kinetic,* while the innervation controlling the entire series of postures, both actual and potential, is *static*. In this light two streams of innervation flow concurrently and in harmony through the production of any purposive motion, the kinetic stream producing movement, and the static stream maintaining actual and potential posture. One or both of these currents may be subject to excessive innervation and produce excessive muscular activity in motion or in posture. Bradykinetic analysis indicates that both types of innervation are excessive in the dystonias with resultant extreme degree of movement and extreme postures amounting in many instances to actual distortions. In effect, therefore, dystonia is due to hyperkinetic and hyperstatic innervation. This is seen in this patient whose trunk tended to a rotary action about the pelvis while he was suddenly raised upon the toes of the right foot and thrown, as it were, as from a springboard. The extreme pronation of the upper right extremity gave the characteristic pose which in the more advanced stages assumes fixation. The torsion of the pelvis, commonly discribed in this disease, was only suggested in this patient, though a study in slow motion of the long muscles of the back and those about the pelvis showed the tendency to rotary motion.

In the study of the individual muscle groups by bradykinetic analysis, those of the right thigh stood out with abnormal intensity during periods which demanded their function only to a slight degree, thus indicating the hyperkinetic innervation. This appeared as a prolongation of muscular action. To a less degree this was conspicuous in the left thigh. It has been suggested that this latter may be an incipient phase of the characteristic deformities in the dystonias. The patient made every effort to overcome the deformity of movement, and in the early stages of this disease there seems to be no question that volition exercises some beneficial influence. Indeed, in a case seen in its incipient stages by one of us (Goodhart) and shown at the New York Neurological Society, exercises, relaxation and re-education added their measure of influence in causing the deformities to disappear entirely. The patient occasionally, and only when extremely tired, shows a momentary dystonic spasm. In the advanced stages, however, voluntary effort at control intensifies the deformities of motion. (Fig. 6.)

2. *One Sister. Dystonia Movements More Extensive and Intense.* The patient, an intelligent girl of eighteen, was the second in this family of three to become a victim of dystonia. Her condition represented a degree of involvement between that of the slightly affected brother and the more advanced case of another sister. There was no retardation in early development, the three children of this family having begun to walk and talk before the age of two.

When twelve years old the patient noticed a sudden jerking or forced movement in the form of pronation, beginning in the right hand. This motor difficulty showed definite advance in the performance of skilled movements generally, particularly in writing, and there were peculiar stiffening and worm-like movements in the fingers which seemed to extend suddenly upon attempts to grasp objects. The left hand soon became similarly affected. Within a few months the right leg showed some disturbance of motility. Gradually a feeling of tension developed in the leg, the extremity became flexed and the hip more or less fixed in this position (Fig. 7). As in all of these cases of dystonia musculorum deformans, this patient became easily exhausted upon attempts at voluntary movements. This is doubtless due in large measure to the efforts at suppression and the extra endeavor to use muscle groups, some of which are already involved. The relation of the trunk and pelvis was such as to cause lordosis of the lower dorsal and lumbar spines, and the entire body assumed an attitude of emprosthotonos. The characteristic hyperextension of the foot was present and a limp much more pronounced than in the brother. There was a marked likeness in the poise and position of the three members of this group. The appearance of all of them warranted Oppenheim's synonym for the condition—*Dysbasia Lordotica Progressiva.*

It is apparent that changes in the static mechanism caused definite alterations in the muscle groups, especially in the hamstrings. Fixation developed at the knee and the hip. The hyperstatic condition was also seen in the inward rotation of the knee as well as in the marked plantar flexion of the foot and the toes. When studied by means of slow motion pictures the hyperkinetic overflow became evident in the soleus gastrocnemius groups. Fixation at the hip restricted the extension so that the trunk was not projected to the usual degree and in the characteristic manner. (Fig. 7.)

3. *Second Sister. Condition More Advanced. Dystonic Movements More Extensive.* The third case of the series was the most advanced of the three children in this family. The patient was a young woman twenty years of age. Her condition was the result of seven years of progressive development. The manifestations began in the lower extremities and gradually extended to the upper extremities. The case is, perhaps, unique, in that it refutes the claim of most observers that the muscles supplied by cranial nerves are not involved in the disease. The facial muscles and those of the tongue were affected to such a degree as to make the patient's speech difficult and almost unintelligible. The speech was of a drawling character, articulation being finally accomplished after efforts to overcome a pronounced hypertension, evidently caused by a hyperkinetic overflow. The extreme muscular activity seemed to be due to sustained tonic spasms in a large number of muscle groups many of which were not pertinent to the particular motor act in process. These spasms produced distortions of the body with marked deformity of motion.

At the usual cinematographic rate of reproduction the patient bore a strong resemblance in her gait, attitude and posture to the brother and sister. There was a similar flexor fixation at the knees, with an equinus fixation of the feet when the patient stood. In locomotion the right heel failed to reach the floor. The typical pronation position of the forearms was emphasized.

In slow motion pictures the analysis discloses the typical hyperkinetic and hyperstatic disturbances. The marked overflow of innervation is apparent in nearly every group of muscles in the body, producing needless and often untimely postures as well as unnecessarily active motions and movements wholly unnecessary to the acts performed. (Fig. 8.)

VI. ANALYSIS OF A CASE OF DYSTONIA MUSCULORUM DEFORMANS SHOWING EXTREME SUSTAINED SPASMS

The patient was a young man, twenty-five years of age, born in Switzerland. His family and personal histories were unimportant medically. The first indication of his malady appeared during his twelfth year when the muscles of the right leg began to act peculiarly, stiffened up and produced an awkward gait. In locomotion, as the condition advanced, the whole body would be brought forward with a "pitch or lurch" to the right.

An important feature in the history of this case was the fact that, following a long period of rest and relaxation treatment, there was pronounced improvement in the dystonia, and for several years there was almost complete cessation of abnormal movements. During this period, under conditions of intense emotion only would there be mild recurrences. From his seventeenth year, however, after an experience causing intense mental strain, there occurred a rapid generalized involvement of the somatic musculature, reaching a remarkable degree of distortion.

At the normal rate of cinematographic reproduction he appeared as a trained athlete, his muscle groups standing out prominently in their unusual development. The hypertrophy was real and the muscle strength in proportion. Testing the muscle capacity by feats of strength it was easy to determine that good muscle power was present, but the lack of proper control made it impossible for him to direct his energies to accomplish any purpose. In endeavoring to wrestle, the patient's opponent felt the full power of the groups of muscles, but the static dissociation threw the patient into involuntary attitudes which defeated his efforts and thwarted his skill. The kinetic energy appeared to develop good muscular power, but produced sustained spasms which caused perversions in all of the motor performances which he undertook. The trunk muscles were quite as much involved as those of the extremities. They threw the body forward and were augmented in their unchecked force by the upward projection of the extensors of the thigh, leg and foot. The force in extension movements was so great as literally to hurl the patient's body forward. In the previous cases attention has been called to the typical equinus tendency which forced the patient to walk upon the ball of the foot. The sudden and uncontrolled flexor and torsion spasms of the trunk with the extension of the arm, gave the impression that a heavy object was about to be hurled with great force. The incessant and powerful muscle activity with its violent contortion suggested a sudden overflow of kinetic energy causing extension of the entire body musculature. Now the flexor groups were in dominant ascending power, and again the extensors. The normally opposed groups seemed to fail in their check function in time to arrest this involuntary overflow.

The details of this extravagant expenditure of energy are revealed in bradykinetic analysis. The characteristic attitudes and poses, clearly showing how the static mechanism gives rise to the clinical pictures, are much more generalized than in any of the previous patients, but the distinguishing features of the muscular deformities are at once recognized. Indeed, if the film is stopped, almost any single picture would enable one to diagnose from its pose the nature of the malady. There is an absence of permanent contractures, conspicuous in some of the other cases, especially those of long duration. This is in part due to the almost universal involvement of the somatic groups, and probably also to the apparent absence of pathological changes in the muscles.

That the emotions exert a strong influence over these bodily movements is generally understood, and this is true of all forms of pyramidal and particularly of extrapyramidal lesions. Sitting in a perfectly quiet environment, the patient had no difficulty in inducing almost complete relaxation, and the abnormal movements ceased. He was able to feed himself and even move about slowly, with almost normal control. Being morbidly sensitive, he felt his isolation to a degree which had a decided effect upon his psyche. The slightest suggestion that he afforded amusement to others intensified his loss of control. A mental attitude developed in this case which tended to bring about an unfortunate state of conflict between his psychic state and his environment. This was not the fact in other cases. It makes necessary a further study of the psychology of the individual, especially in relation to motor control. (Figs. 9 and 10.)

VII. ANALYSIS OF A CASE OF DYSTONIA MUSCULORUM DEFORMANS SHOWING THE "DROMEDARY ATTITUDE" AND SO-CALLED SEMILUNAR FOOT

The patient was a girl of seventeen with good personal and family history. The long duration (ten years) and extreme degree of the manifestations peculiar to this disease give the case an especial interest. Born in this country of Russian Jewish parentage, the girl's first seven years of life showed normal development. At about the age of seven, peculiar, involuntary movements appeared, which were at first limited to the right foot but gradually spread to the right hand and arm. The description of these initial motor symptoms given by many of the patients suffering from this type of the disease, furnishes a conception of the character of this muscular involvement. It suggests what is understood under the term spasm.

In the course of the following three years various muscle groups became affected while involvement of the musculature of the back and neck was followed by that of both extremities upon the left side. The extension of the disease to all of the somatic groups covered a period of from three to four years, culminating in the incessant movements characteristic of her condition. The muscles supplied by the cranial nerves, with the exception of the sternocleidomastoids, and the trapezii were not involved. When in the prone position, the patient found lying on the left side most comfortable and the extreme deformities, those that were transient and those that were permanent, were then well demonstrated. The patient's movements, because of contractures and resulting deformities, were extremely slow, so that the contrast between normal and high speed photography is not so striking in this case as in others. The appearance of the patient in the cinematographic reproduction at normal speed was that of the typical *dromedary attitude and locomotion*. The extreme deformities of the feet were unusual, and had it not been for a definite history, we would have suspected a condition of congenital clubfoot.

In the slow motion pictures the characteristics of dystonia are demonstrated, though the extreme contractures and joint deformities have changed the original attitudes and now limit the play of muscular groups. Even in the normal projection and now more clearly analyzed, the movements and postures appear as overaction of certain groups of muscles, and it can be readily seen why the term dystonia has been adopted. As a matter of fact, the defect is due to hypertonia, inasmuch as the hypotonia is relative and not real. However, lordosis and dysbasia are even more obtrusive symptoms in nearly all cases. There is a fixed flexion at the hip and knee, with inversion and extension of the right foot, while the left foot is extended. The phalanges of the feet are strongly flexed increasing the marked concavity present in the plantar arches, and giving rise to the formation of what is called "semilunar" foot. So extreme are the deformities that the plantar surfaces of the feet do not touch the floor. Closer study brings out flexor-extensor movements of the fingers of the paralysis agitans type. Abnormal involuntary movements in various groups of muscles keep the body, both in the prone and the upright position, in continuous action. The muscular contractions of the various groups are so constant as to keep up a series of sustained, arrhythmical spasms. The muscles of the lower extremities were atrophied while those of the upper extremities and their axio-appendicular groups were greatly hypertrophied. The partial atrophy of the muscles of the legs was doubtless due to local changes in the joints, those of the ankles and feet being ankylosed. These were doubtless secondary mechanical results of the muscular disorganization.

Sudden tic-like movements of muscle groups in the upper extremities appeared, occasioned when the patient was under emotional strain. They appeared to be myoclonic in type. In no single case of dystonia have we seen such a combination of abnormal movements. Indeed, one might hesitate to designate this case as one of pure dystonia musculorum deformans, although it possessed the distinctive features. (Fig. 11).

VIII. ANALYSIS OF A CASE OF DYSTONIA MUSCULORUM DEFORMANS WITH PROGRESSIVE IMPROVEMENT

This patient is one of those cases of dystonia which have shown improvement by means of physical re-education. Bradykinetic analysis reveals certain deformities of motility which our previous studies have shown to be characteristic of the organic affection. The boy was normal until his eleventh year. About a year previous to this he fell from a fire-escape on the first floor of a building. He landed upon his feet and feeling no ill effects whatever, continued playing. There was nothing in the history of the following year to indicate any connection between the fall and the onset of his illness one year later. The first symptom was an outward rotation of the right lower extremity. Some two years later sudden involuntary pronation movements of the right hand, followed about a year later by an involvement of the left lower extremity, completed a picture which gave the patient the characteristic cast of the milder type of dystonia. His appearance on re-examination showed a decided improvement as compared to that of about a year previous. Nevertheless, viewed in the light of the patients already studied, there was characteristic pose and attitude of the body as a whole, and of the extremities in particular.

In the slow moving pictures the right lower extremity showed peculiar external rotation at the hip, knee and ankle, with hyperextension of the foot in sustained equinus position. Bradykinetic analysis of this motor phenomenon was, in itself, sufficient to make the diagnosis. The tendency to pronation position of the left upper extremity; the extreme extension of the toes of both feet at the same moment as the patient was about to turn, the pronounced upward shift of the entire right lower extremity, the lordotic curve with the suggestion of torsion in the trunk, were unmistakable.

The patient was next observed as he moved "on all fours." This position, as the patient moved forward, brought out even more emphatically the special deformity apparently due to defective postural fixation. Obviously, this deformity of movement would not have been apparent had he moved forward on hands and knees; the defect in attitude just alluded to would not have been brought out, since the extensors of the lower leg and foot would thus not have been brought into play. To demonstrate this fact it was necessary to observe him moving forward resting on fingers and toes and ball of foot. (Figs. 12 and 13.)

In summary, the analysis of these cases of dystonia musculorum deformans reveals the fact that an actual disorganization exists in the relation of the static and kinetic mechanisms. The marked hyperstatic defect appears to produce initial and intermediate postures of such extreme fixity that an unusual amount of kinetic energy is required to overcome them. The dystonia, therefore, seems to arise from a marked imbalance in the ratio of static energy to kinetic energy. This is particularly true during volitional effort. The pronounced hyperkinesis during inaction indicates an unstable condition in the kinetic system, more probably in that portion concerned with automatic associated movements, for the reason that the motor disorders are suggestive of movements of this kind. The disturbance in both the static and kinetic mechanisms partakes of the characteristics indicative of irritation. In addition to the evidence of irritation in these two mechanisms, the analysis points to a disturbance in the rhythm and interrelations of these mechanisms, which, being improperly synchronized in time, force and rate of action, lead to a conflict between hyperstatic and hyperkinetic innervation.

IX. ANALYSIS OF A CASE OF CEREBELLAR ATAXIA

The patient was a man forty-one years old whose defects in motility were first observed in infancy. Voluntary efforts of upper or lower extremities, or of the trunk in change of body posture, were always attended by irregular oscillating movements.

His equilibratory defects made locomotion difficult. Incoordination in the speech mechanism imparted a jerky quality to articulation which in consequence was often unintelligible.

At the cinematographic rate of reproduction, the patient demonstrated a marked ataxia in extremities and trunk. The slow motion pictures furnished opportunity for the analysis of the disturbance. When the patient attempted to walk he suddenly lost his equilibrium and fell. As the trunk approached the ground, and an effort at balance was made by the patient, the normal interplay of agonists and antagonists was seen to be defective. The segmental portions of the body as it fell did not follow in sequence nor did they retain their relationship of posture and attitude as seen in the normal. There was evident loss of associative static control. The same group dissociation was seen upon studying the efforts to rise. The extreme disproportion in muscular effort was due to lack of harmonious action between muscle groups. As he attempted to regain the upright position, he sustained for a moment his entire body weight upon his hands; taking advantage of the fact that there was no reduction in dynamic force, he attempted to hurl his trunk upward, thus to attain the upright position without going through the normal sequence of segmental attitudes. With the usual cinematographic speed the details of these pictures would have been beyond visual recognition.

A study of the details of defective motility not associated with equilibration and yet dependent upon synergic control was offered by a series of pictures showing the finger-to-nose test. The hand and finger on approaching the nose showed irregular oscillations which gradually increased toward the end of the act. The oscillations in the upper extremity were due to lack of balance between the components of the synergic units of the arm groups. In analyzing the nature of the rebound phenomenon of Gordon Holmes, in slow motion projection, the wrist when suddenly released by the examiner, rebounded forcibly striking the cheek. This rebound was due to a failure of the check component—the triceps in this case—to play its part properly in the synergic unit during flexion.

A projection study of the motor phenomenon of the pendular knee reflex shows that this is also due to an inadequate relation between the check and dominant elements in the synergic units producing flexion and extension of the leg on the thigh. Normally the impulse conveyed by tapping the extensor tendon stimulates the quadriceps extensor, the dominant element, and the posterior thigh group, which act as the check element. Working as a synergic unit the two groups during the production of the knee-jerk restrict the motor reaction to one to three oscillations of the leg. This is due to the mutual check action of the two muscle groups affected by the reflex. This fact has been demonstrated (Tilney) by kymographic records in the case of all normal reflexes when, as in this case,·the synergic unit for extension and flexion of the leg is dissociated, the mutual check action is lost and this pendulum-like swinging results.

A striking feature of the bradykinetic analysis in this case was the long interval of time that lapsed between the application of the stimulus and the response as seen in the knee-jerk. As the hammer slowly descended upon the tendon of the quadriceps extensor there was a comparatively long interval of latency before the leg began to move.

In summary, the analysis of this case of cerebellar aplasia reveals the fact that the ataxia is due to a failure of the proper coordination in the synergic units of the body and that the disproportionate innervation of the dominant and check elements in each synergic unit leads to the characteristic disorder of cerebellar lesions, *asynergia*. (Figs. 14, 15, 16.)

CONCLUSIONS

1. Bradykinetic analysis by means of slow motion pictures furnishes one of the most direct approaches to a clinical study of disturbances of motion.

2. Adventitious elements in motor dis-

turbances are manifest as disorders either in the kinetic or static mechanism.

3. Cinematographic analysis indicates that all purposive movements depend upon the concurrent and synchronized operation of the kinetic and static mechanisms. Such movements are made up of a series of postures beginning with an initial posture and concluding with a terminal posture. This series of postures during a movement may readily be demonstrated by stopping the projection apparatus at each picture either in the slow or normal reproduction. Such an act as taking a step forward then appears to be composed of a series of successive postures.

4. The kinetic mechanism is represented by a neural apparatus capable of correlating the impulses necessary to the production of purposive movements, attitudes and postures, ultimately giving these their proper motor expressions. It, so to speak, prescribes and produces movements and postures for which reason it is a *kinethetic* (causing motion) system.

5. The static mechanism is represented by a neural apparatus capable of correlating the impulses necessary to the proper maintenance of postures. Ramsay Hunt has graphically illustrated the relation of posture to motion in his expression that "movements are followed by their attendant postures like shadows." Analysis by the slow moving pictures verifies this conception. The static mechanism, so to speak, controls the postures which follow movements like shadows, stabilizing them through all their phases. In this sense and because it maintains posture, it is a *statothetic system.*

6. From such evidence as is at present available, it seems warranted to conceive of a static arc representing the statothetic system with its chief center in the cerebellum, and a kinetic arc representing the kinesithetic system whose centers are in the endbrain. The trend of recent opinion would allocate the primordial center for the kinetic arc (paleokinesis—Hunt) to the corpus striatum or basal forebrain ganglion.

7. The disorders of motion investigated in this study, including choreiform movements in chronic degenerative chorea and astasia abasia, cerebellar ataxia in a case of cerebellar aplasia, intention tremor in multiple sclerosis, and dystonia in dystonia musculorum deformans, may be attributed to disorganizations in the two fundamental neural arcs already described.

8. Choreiform movements in degenerative chorea, upon analysis, appear to be due to a hyperkinetic disorder resulting in the sudden productions of postures and attitudes directly expressive of the emotions of fear, anger, disgust, etc. The static mechanism is affected to a lesser degree as shown in the instability of posture indicating an imperfect association of the synergic units. Choreiform movements in astasia abasia are likewise based principally upon hyperkinetic defects. The resulting disorders of motion produce postures and attitudes expressive of the emotions. In this case these emotions are more highly complex than in degenerative chorea, and indicate a more acutely conscious and psychic elaboration of fear.

9. Cerebellar ataxia and intention tremor, upon analysis, both appear to be due to essentially the same defect limited to the static mechanism. The motor disturbances in these conditions are produced as a result of a disorganization in the proper relation of the synergic units of the body.

10. The dystonias, upon analysis, are clearly revealed as hyperkinetic and hyperstatic in nature. The normal concurrence in these two streams of static and kinetic innervation is imperfect or wanting. These mechanisms appear to be affected by pronounced irritation or lack of proper inhibition.

11. The foregoing suggestions on the basis of bradykinetic analysis are offered with the desire of furnishing an anatomical and physiological background for certain motor disorders and at the same time of calling attention to the seemingly essential relationship of the static and kinetic mechanisms in all purposive motion. Much further study is needed in the development of these conceptions before they may be advanced as more than working hypotheses.

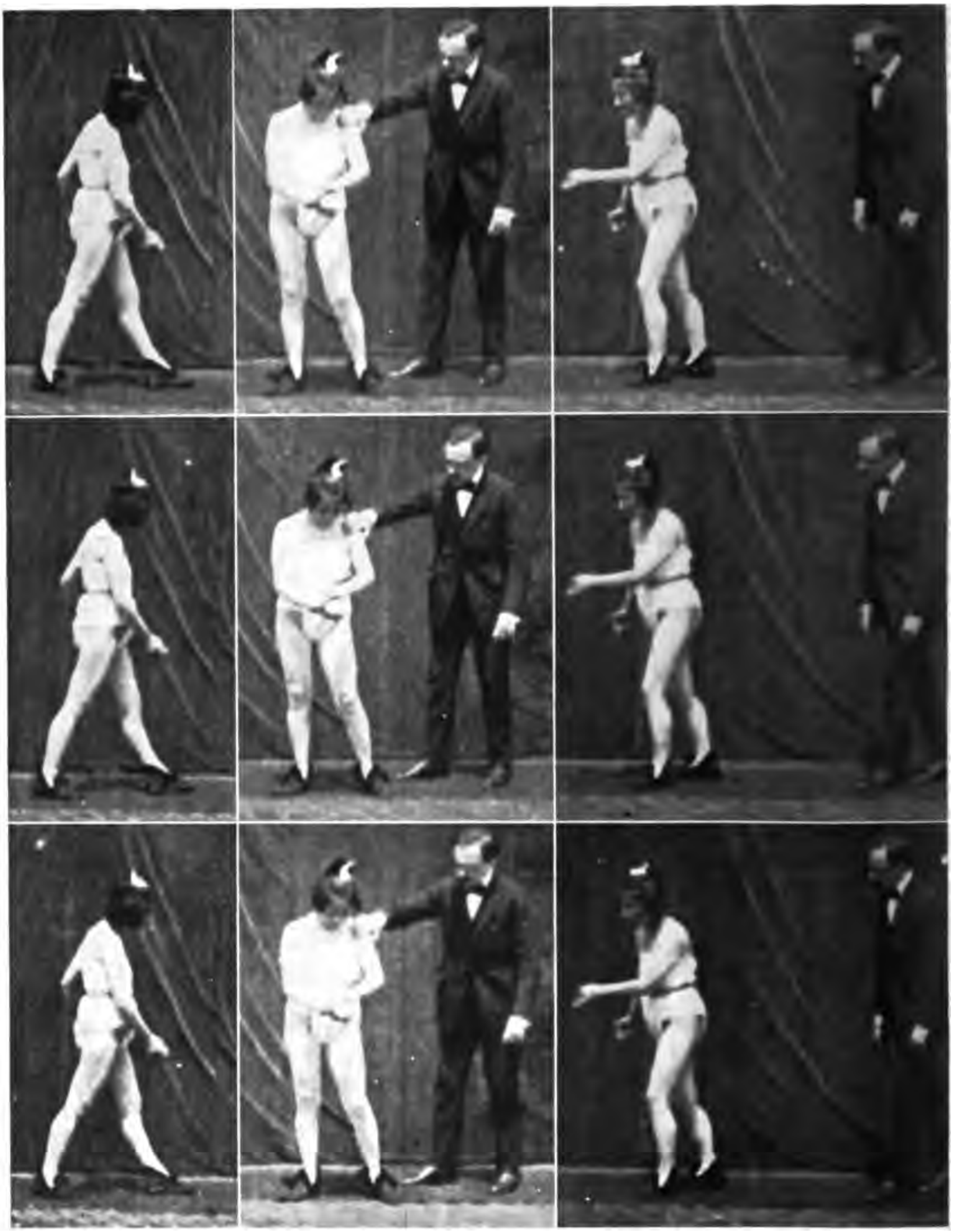

FIG. 1. Bradykinetic Analysis in a Case of Chronic Degenerative Chorea with Athetotic, Choreiform and Attitudinizing Elements.

What appeared by normal projection as purposeless, meaningless movements, by bradykinetic analysis are seen to be gesticulations, expressions of definite acts of going, of giving; and attitudes and gestures symbolizing emotions of shame, coyness, turning away, etc. The hyperextension of the left leg at the knee is seen in the picture. Athetotic (polypoid) movements of the fingers are indicated. Facial grimacing is indistinctly discernible in the photographs though clearly seen on the screen.

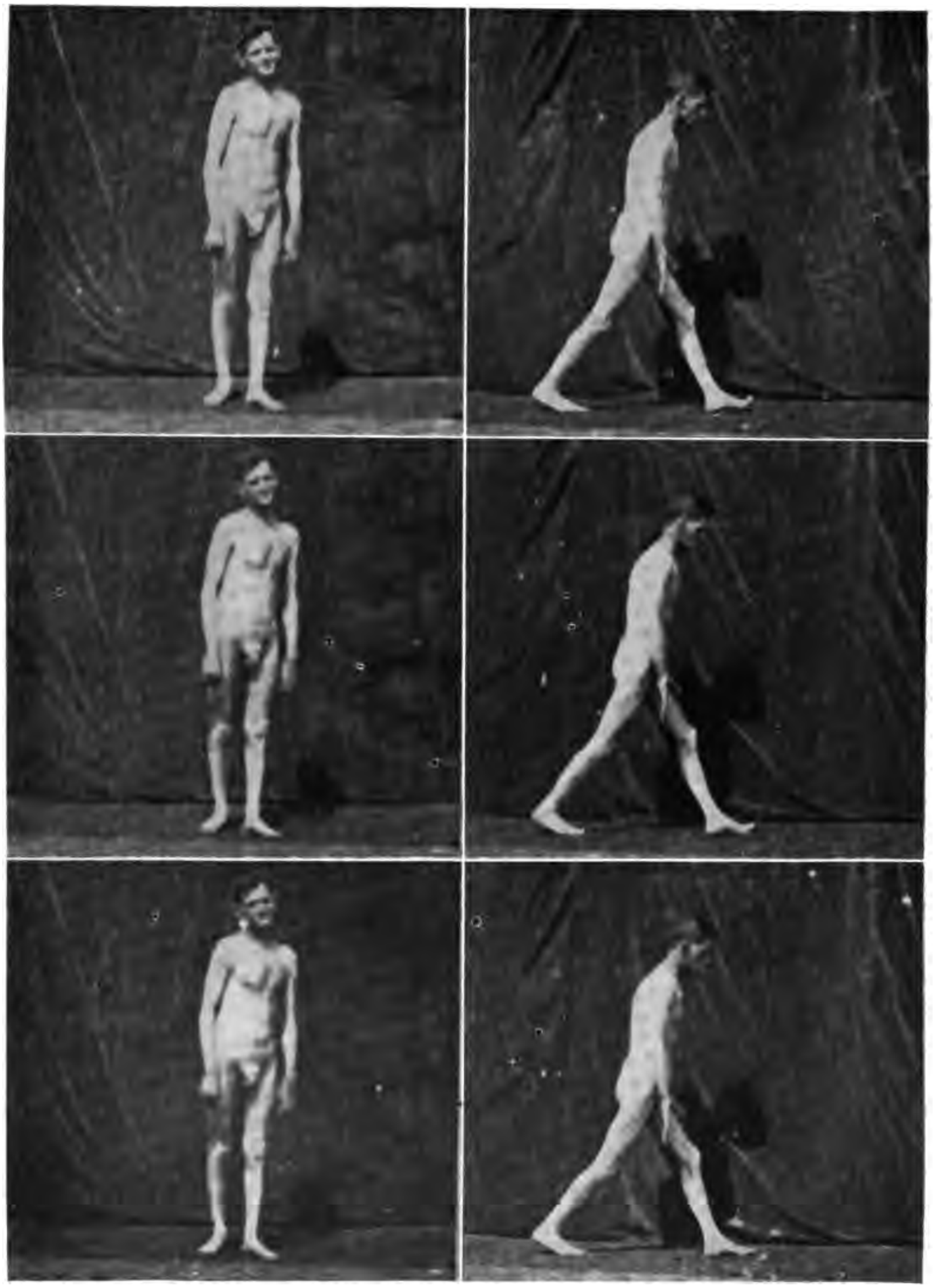

FIG. 2. Bradykinetic Analysis in a Case of Chronic Degenerative Chorea with Some Elements Suggestive of Dystonia.

Torsion movement involving upper trunkal and thoracic musculature: pronator position of hand and forearms. Extensor position of big toe—not a Babinski, however. Overextension of foot in walking, almost a constant feature of gait in dystonia, is not seen in above: on the screen, however, it is occasionally observed in this patient but to a far less degree and inconstant. Some grimacing and continual choreiform movements are of a type present in chronic degenerative chorea. The torsion of the trunk involves rather the upper trunkal segment in contradistinction to that seen in dystonia in which there is pelvo-trunkal torsion.

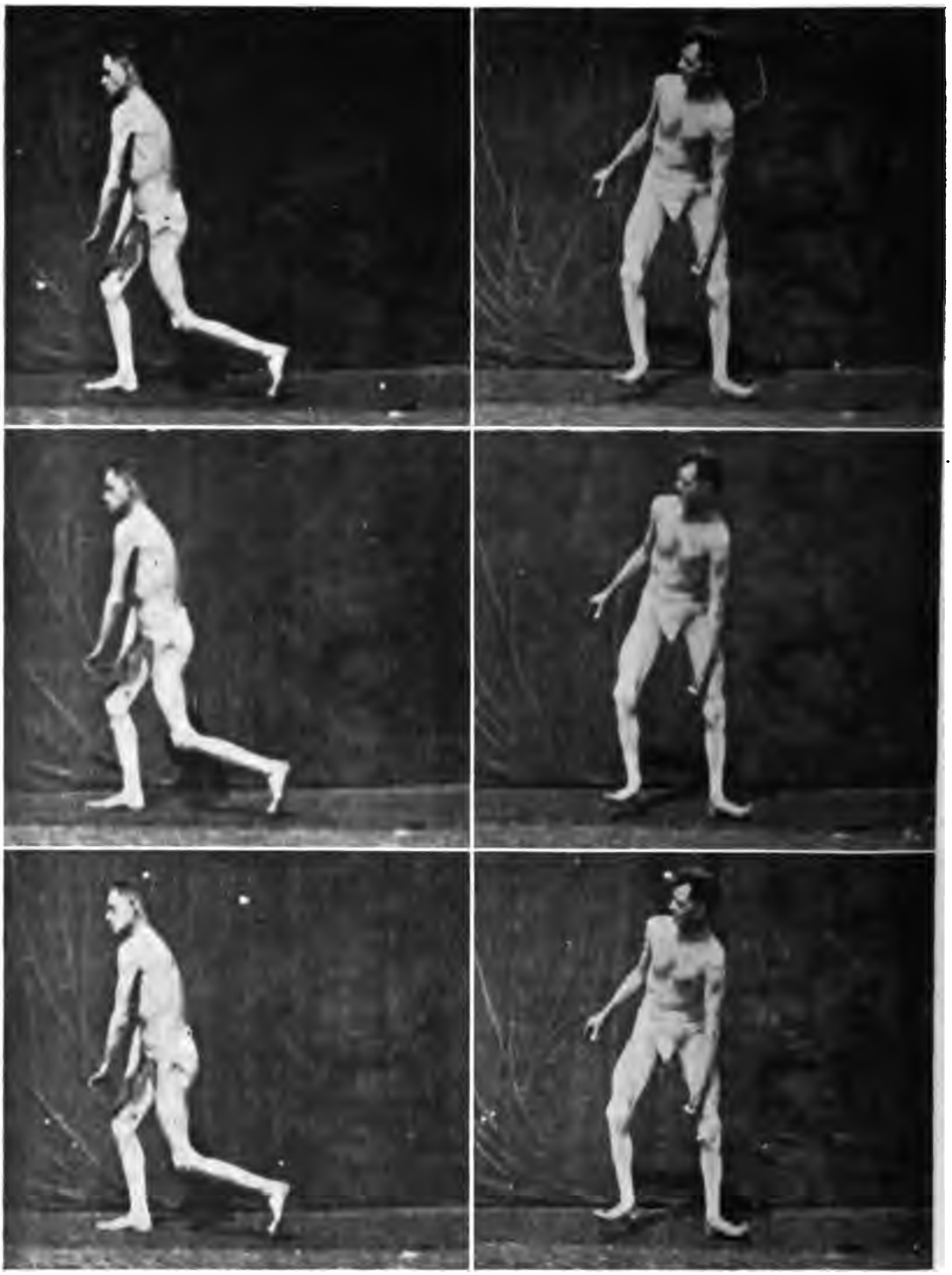

FIG. 3. Same Case as figure 2.

Gait with overflexion at the knees and pronator position of forearm with right arm fixed on thigh during locomotion give dystonic character to attitude; attitudinizing as seen in act of hurling an object. (Compart with figure 9).

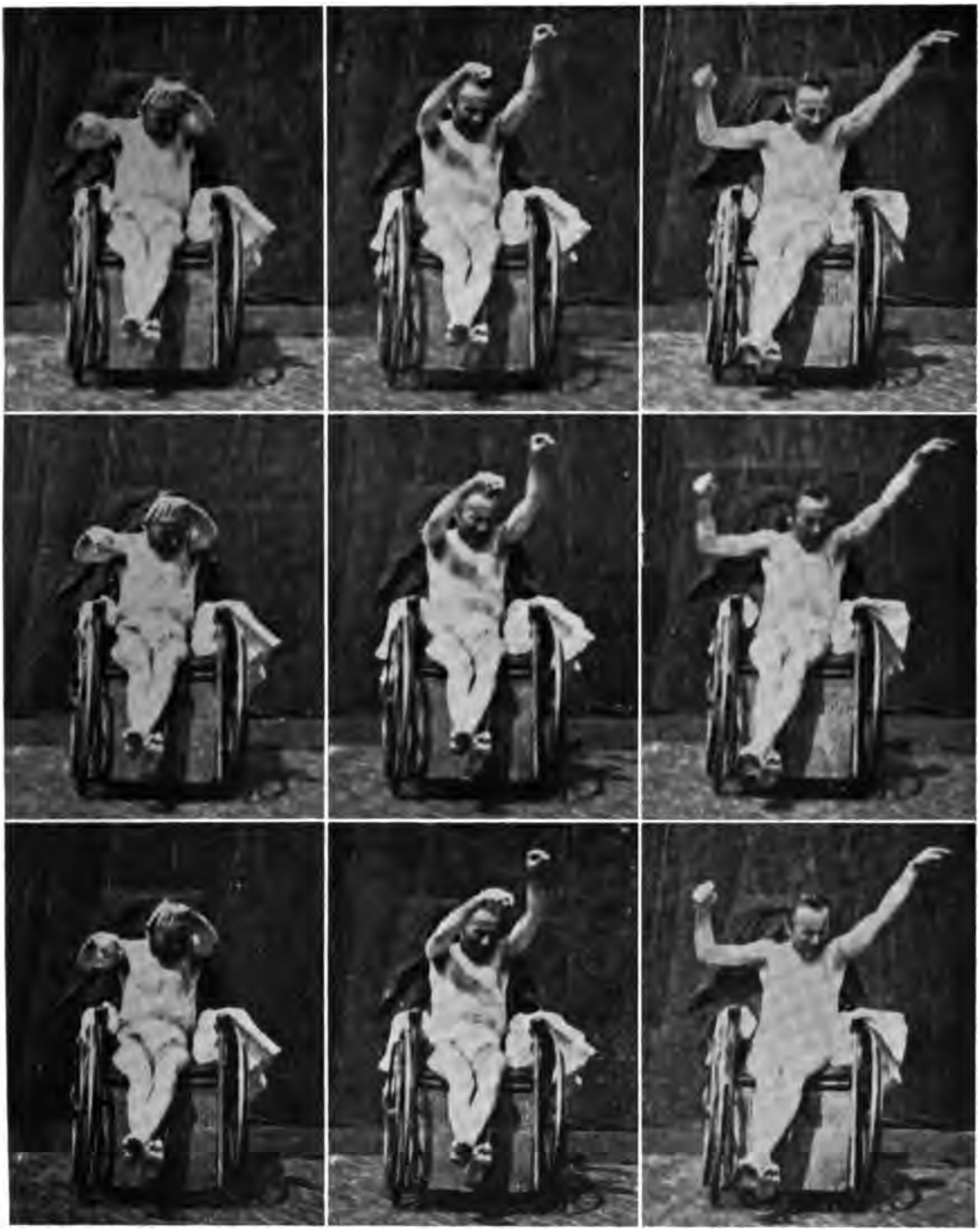

FIG. 4. Bradykinetic Analysis in a Case of Multiple Sclerosis of Dyssynergic Character. Finger-to-Finger Test; Dissociation of Synergic Unit.

Observe dyssynergy even as patient attempts to place his upper extremities in position for approximation of the widely separated digits. The defective morphology of group movement becomes more evident as patient attempts to bring his fingers together. The arms shoot far beyond the point desired, each element of advance showing the character of the deformity of motion, the dissociation of the synergic units. The lower extremities and trunkal muscle groups are all involved, the case being one of so-called integrative asynergia.

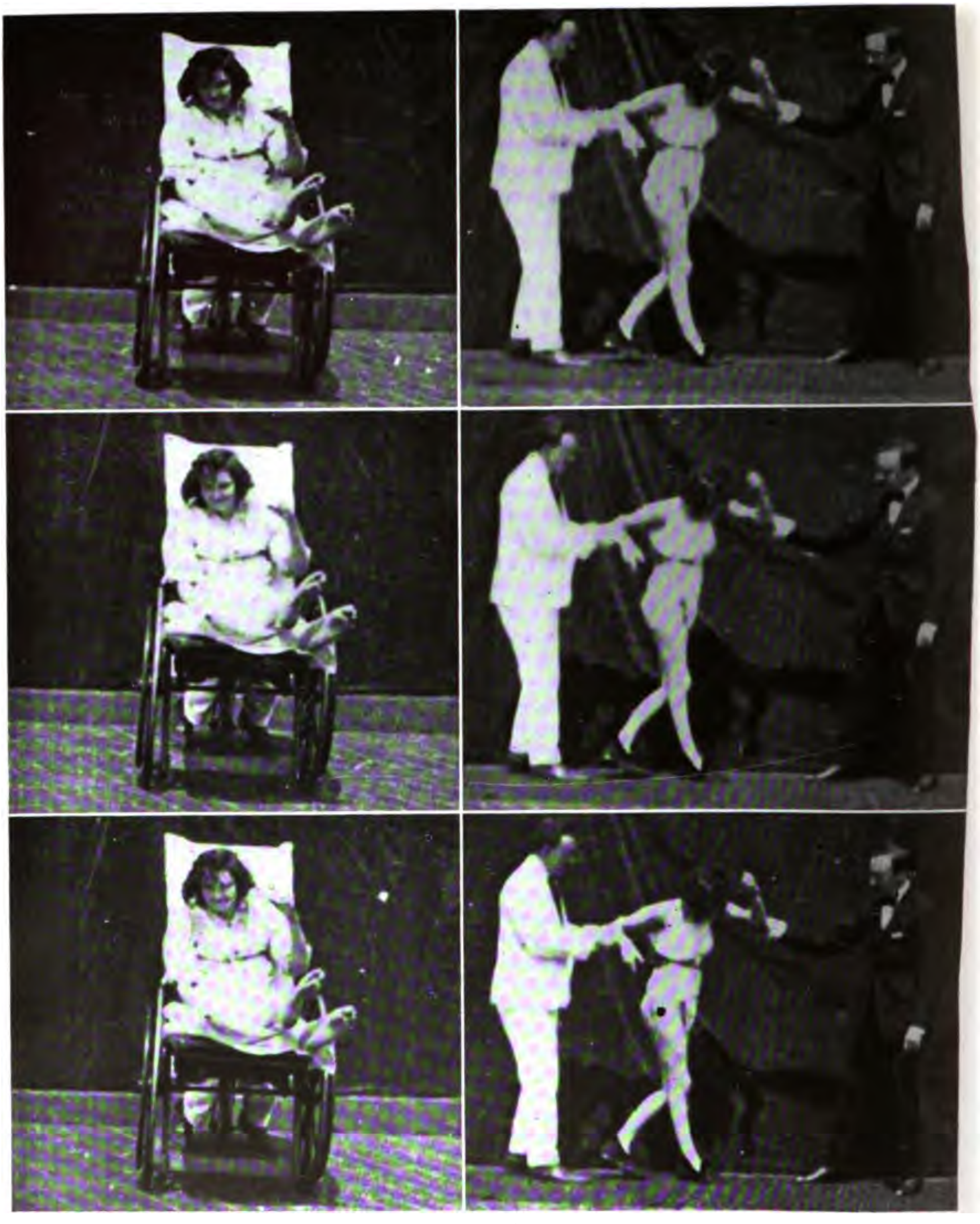

FIG. 5. Bradykinetic Analysis in a Case of Astasia-abasia with Excessive Motor Agitation.

Motor manifestations classically those of psychic origin. With patient seated the foot and toes show tendency to plantar contraction there being no paralysis of extensors and no true contractures of flexors. The upper and lower extremities are in constant movement showing distinctly a pattern; static and kinetic mechanisms act normally and in unison. Upper extremities are engaged in rythmic movements imitative of those of flying. Vermicular wave described in text is not discernible in photograph.

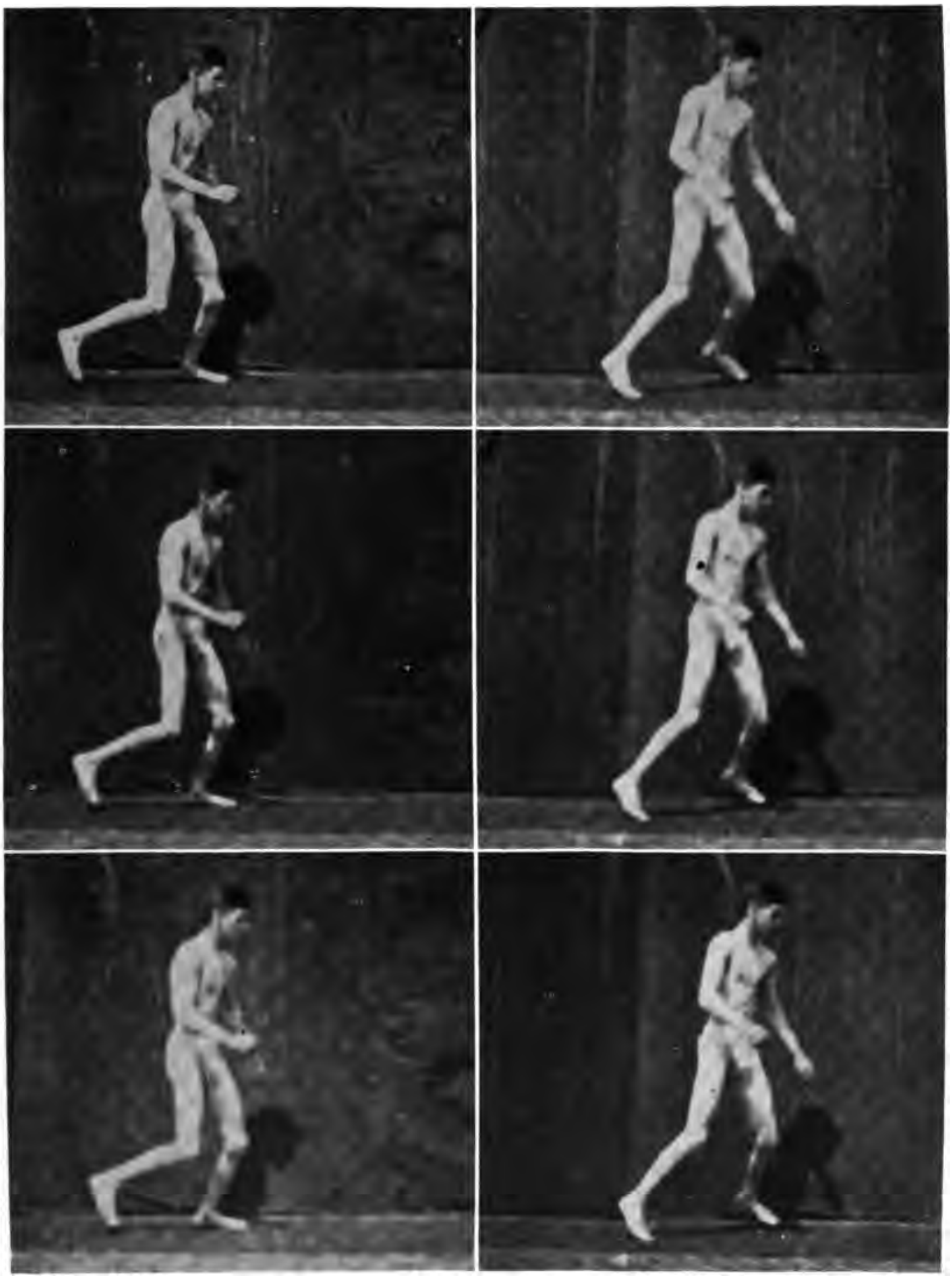

FIG. 6. Bradykinetic Analysis in a Case of Familial Dystonia Musculorum; Group of Three Members of One Family: A Brother and Two Sisters.

The above picture is that of the brother, the last to be afflicted, and showing the incipient stage. Characteristic inward rotation at knee, equinus position, overextension of foot in action with accompanying flexion at hip and knee. Extreme pronation position of forearms and hands not due to contractures.

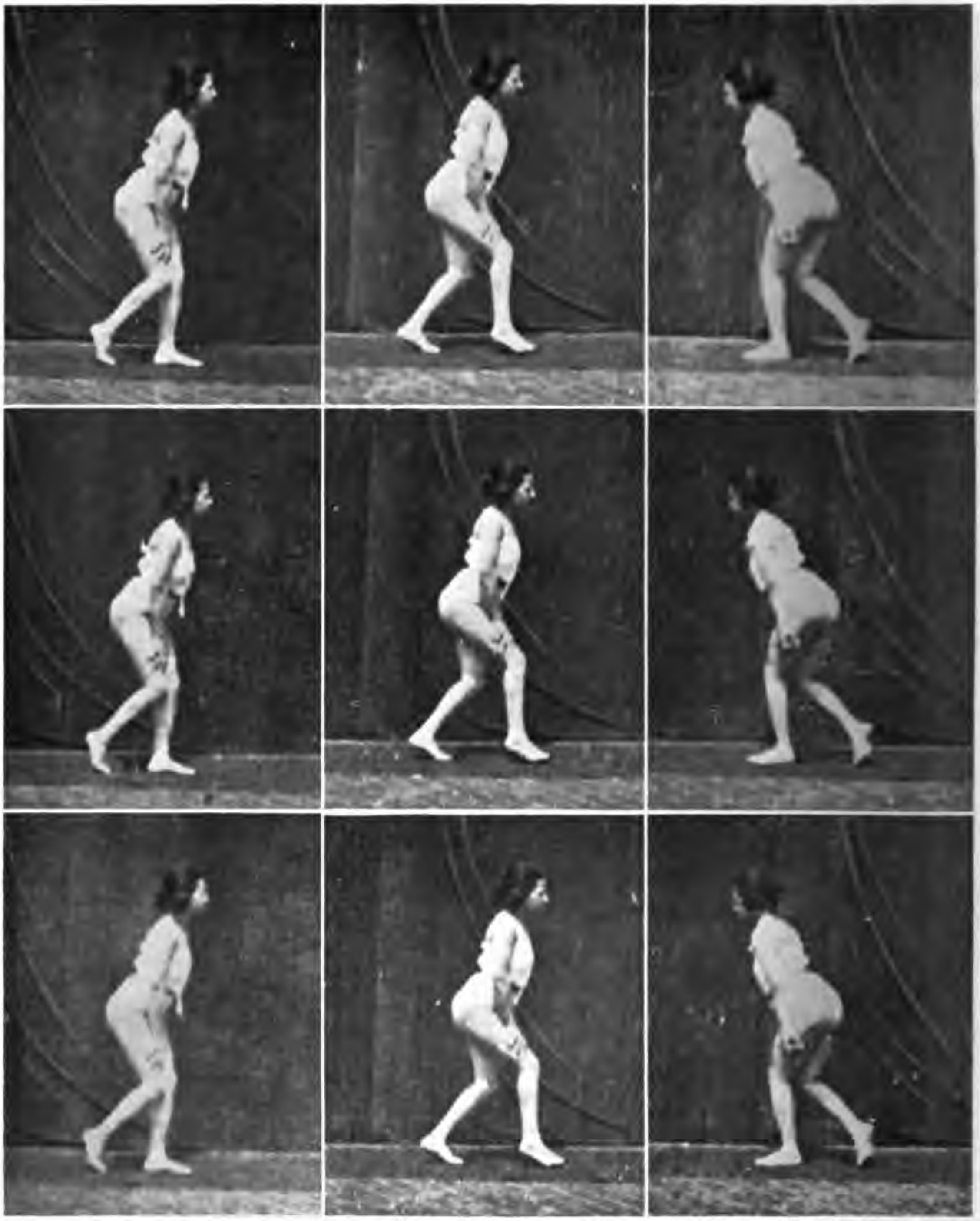

FIG. 7. Bradykinetic Analysis in a Case of Familial Dystonia Musculorum; One Sister.

Symptoms more pronounced than in the case of the brother (preceding picture). Observe lordosis, flexion at hip and knee. Flexure contractures of right hip and knee joints are fixed by muscle changes. Inward rotation knee and hip, also hyperextension at ankle while walking, together with pronator position of forearm and hand are shown in the picture. The mechanism in muscular group action is brought out in the animated series on the screen.

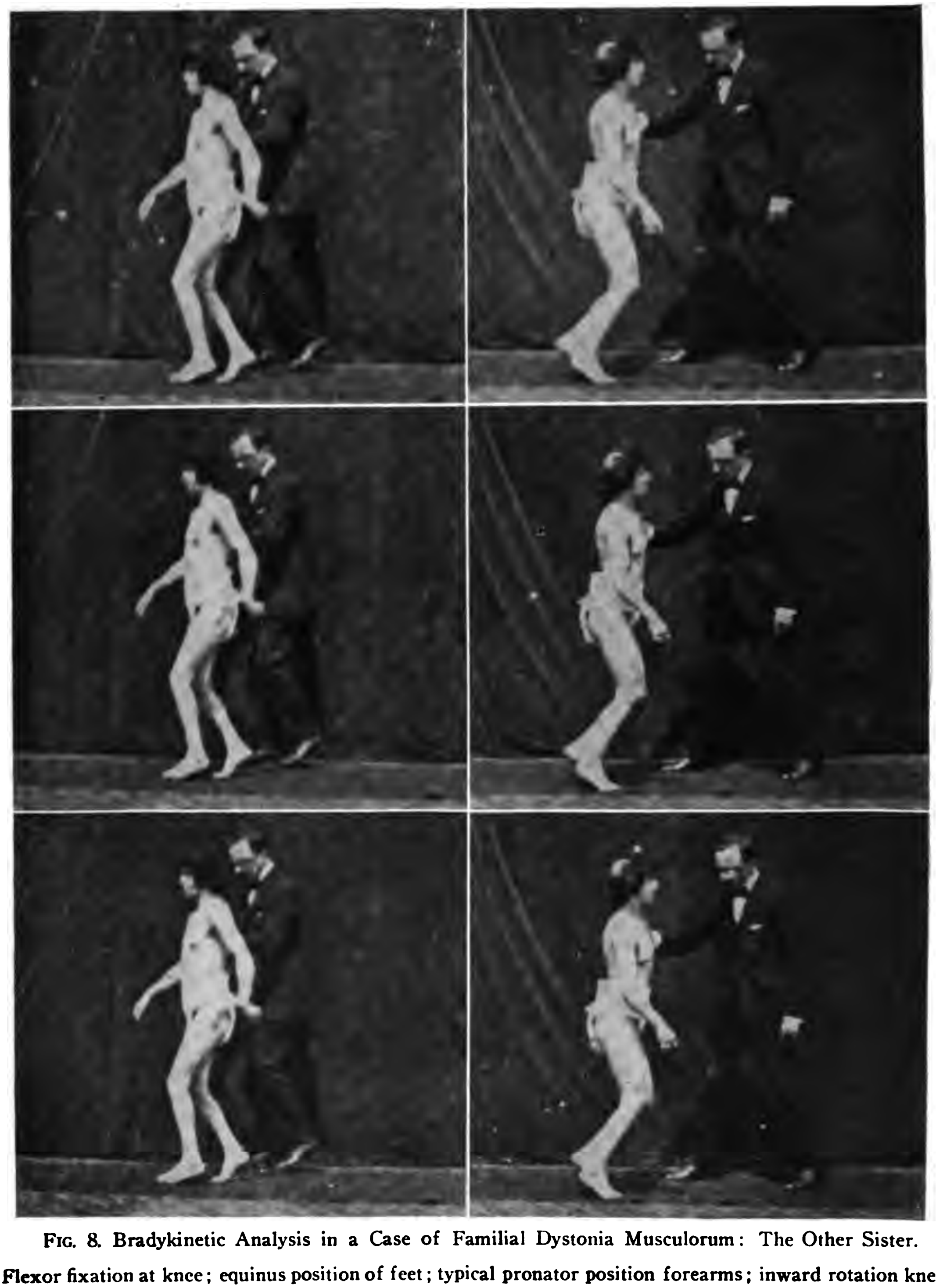

FIG. 8. Bradykinetic Analysis in a Case of Familial Dystonia Musculorum: The Other Sister. Flexor fixation at knee; equinus position of feet; typical pronator position forearms; inward rotation knees.

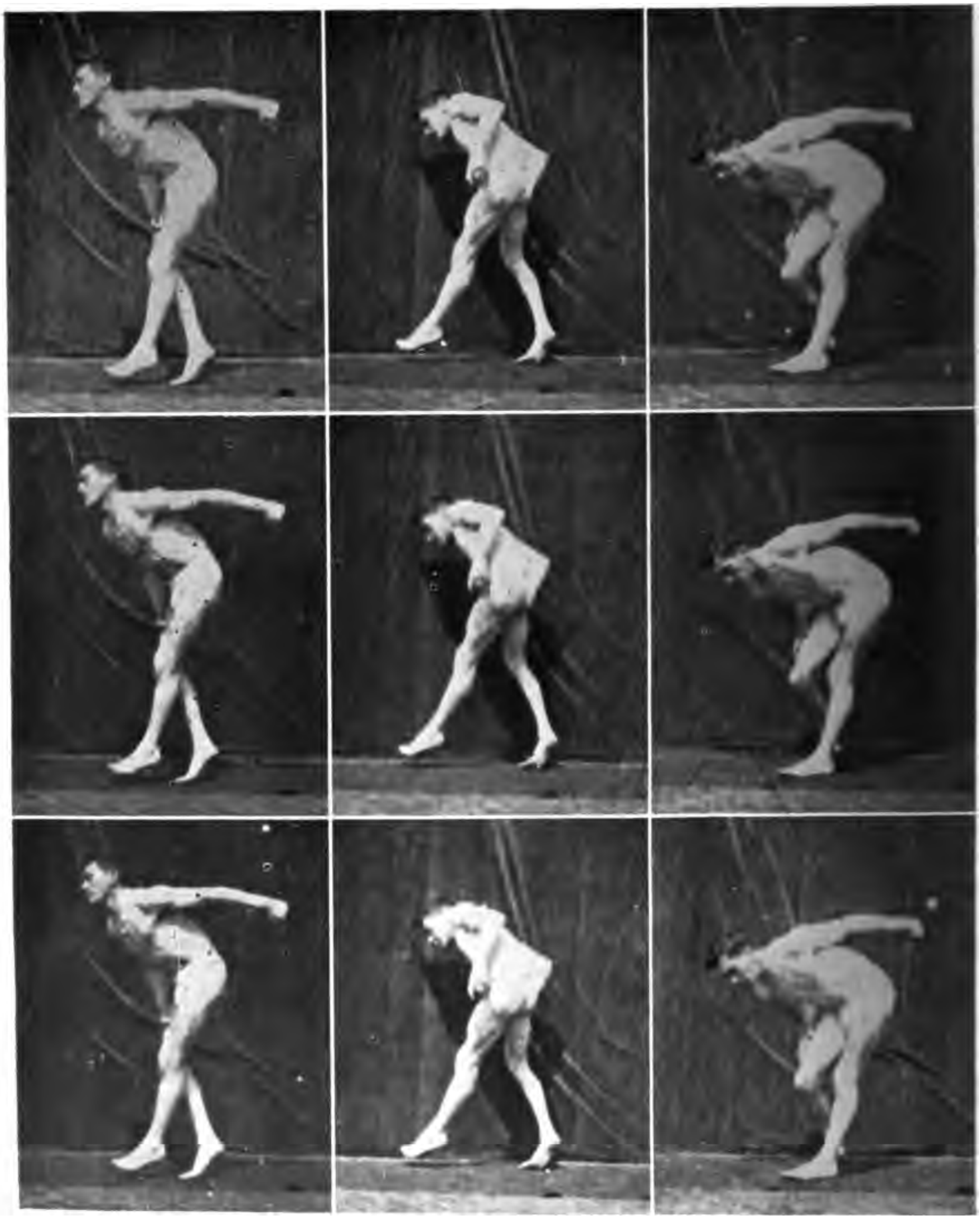

FIG. 9. Bradykinetic Analysis in a Case of Dystonia Musculorum Deformans. Involvement of All Groups of Muscles and True Hypertrophies.

Bizarre distortions of movement. Characteristic positions. Plantar flexion at ankles in walking. In some position inward rotation at knee. Trunk muscles also involved. Body thrown violently forward and movement intensified by upward projection by extensors of hip, knee and ankle. Hand resting on thigh to restrict extreme sudden flexion at hip. Some tendency to pelvic torsion. Force in extension movements so great as to literally hurl the body forward. Sudden uncontrolled flexor and torsion movements give impression patient is about to hurl heavy object. Suggestion of sudden release of kinetic energy not controlled in static fixation. There is no permanent fixation by contractures in this case.

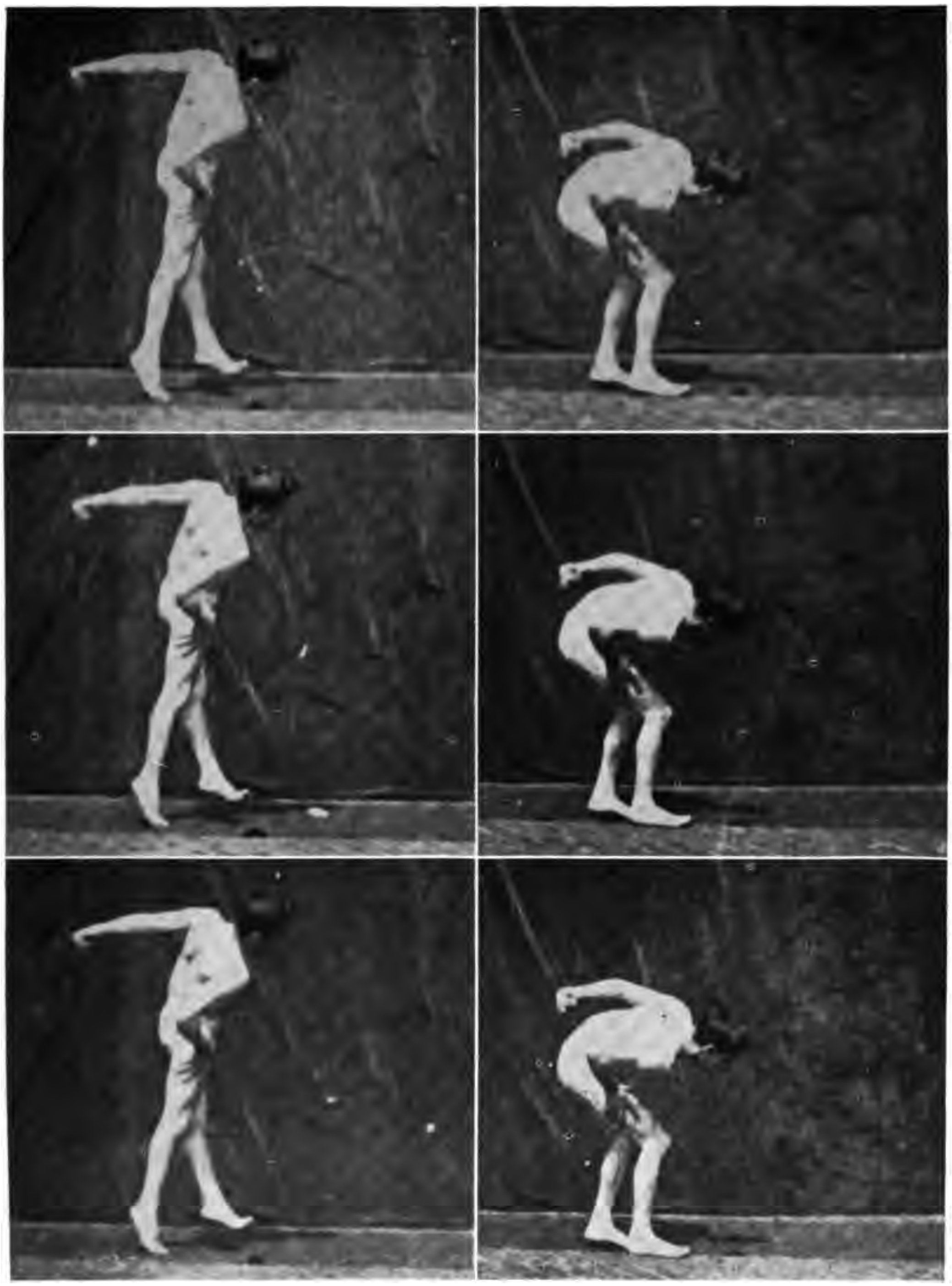

Fig 10. Bradykinetic Analysis in a Case of Dystonia Musculorum Deformans.

Same as figure 9. Observe heer occasional hypexertension of toes, not often seen in dystonia.

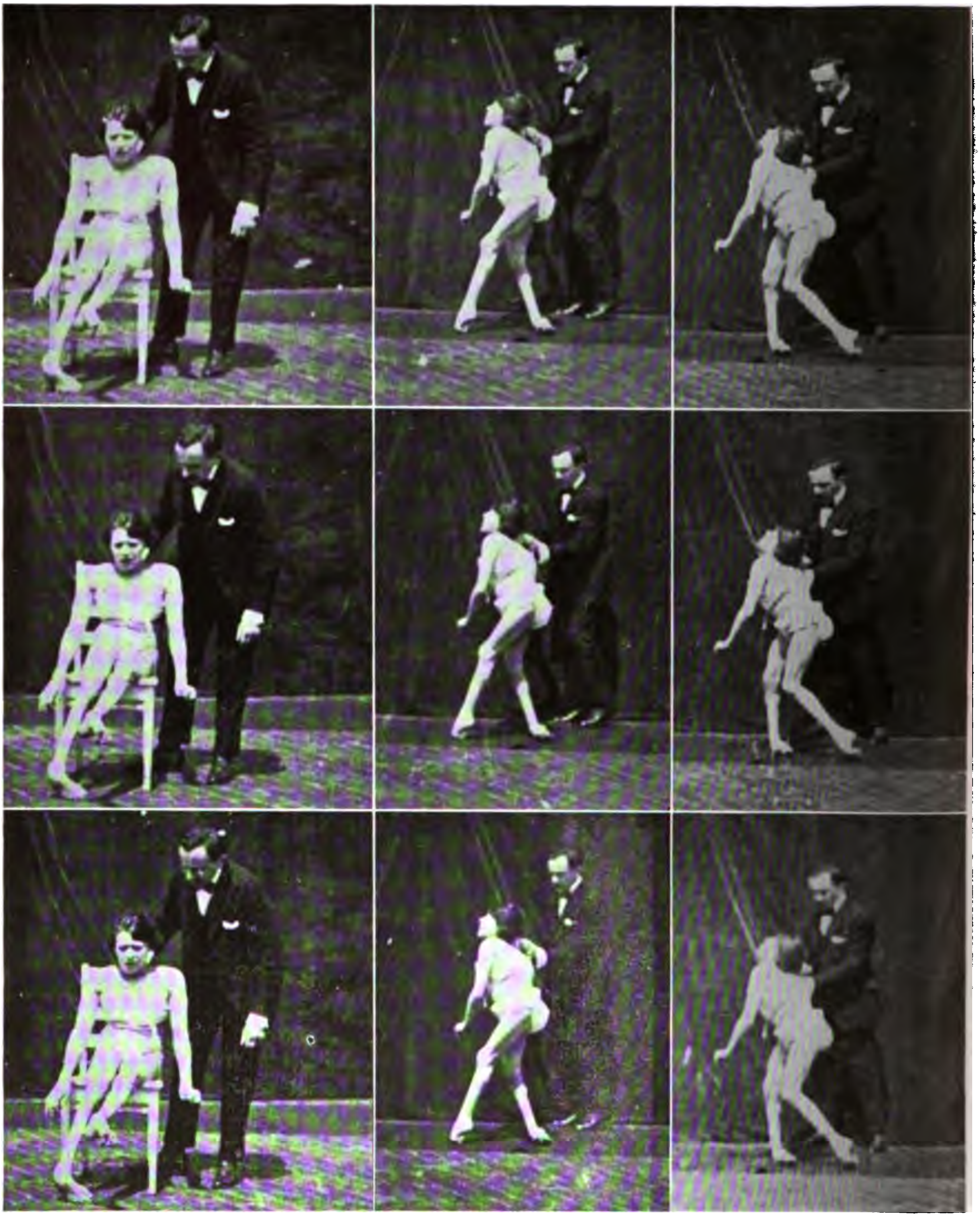

FIG. 11. Bradykinetic Analysis in a Case of Dystonia Musculorum Deformans with "Dromedary Attitude": Semi-lunar Foot.

Extreme deformities of feet; girl walks on dorsal surfaces of feet. Extensor and equinus positions have become "fixed," in less intense degree "fixation" positions in flexion have also developed at hip and knee. Lower extremities below knee relatively atrophic from partial disuse after ankylotic changes though the tibial and calf groups are still in constant myotonic activity as far as the joint changes permit. Upper extremities are in constant motion and all muscles hypertrophied. The trapezii are especially active causing ceaseless clonic extensor movements of the head. There is extreme deformity of trunk due to fixation by domination of certain muscle groups of the trunk and pelvis. Forearms are in characteristic positions. In the moving pictures a distinct rhythm of the paralysis agitans type can be observed in the movements of the forearms and hands.

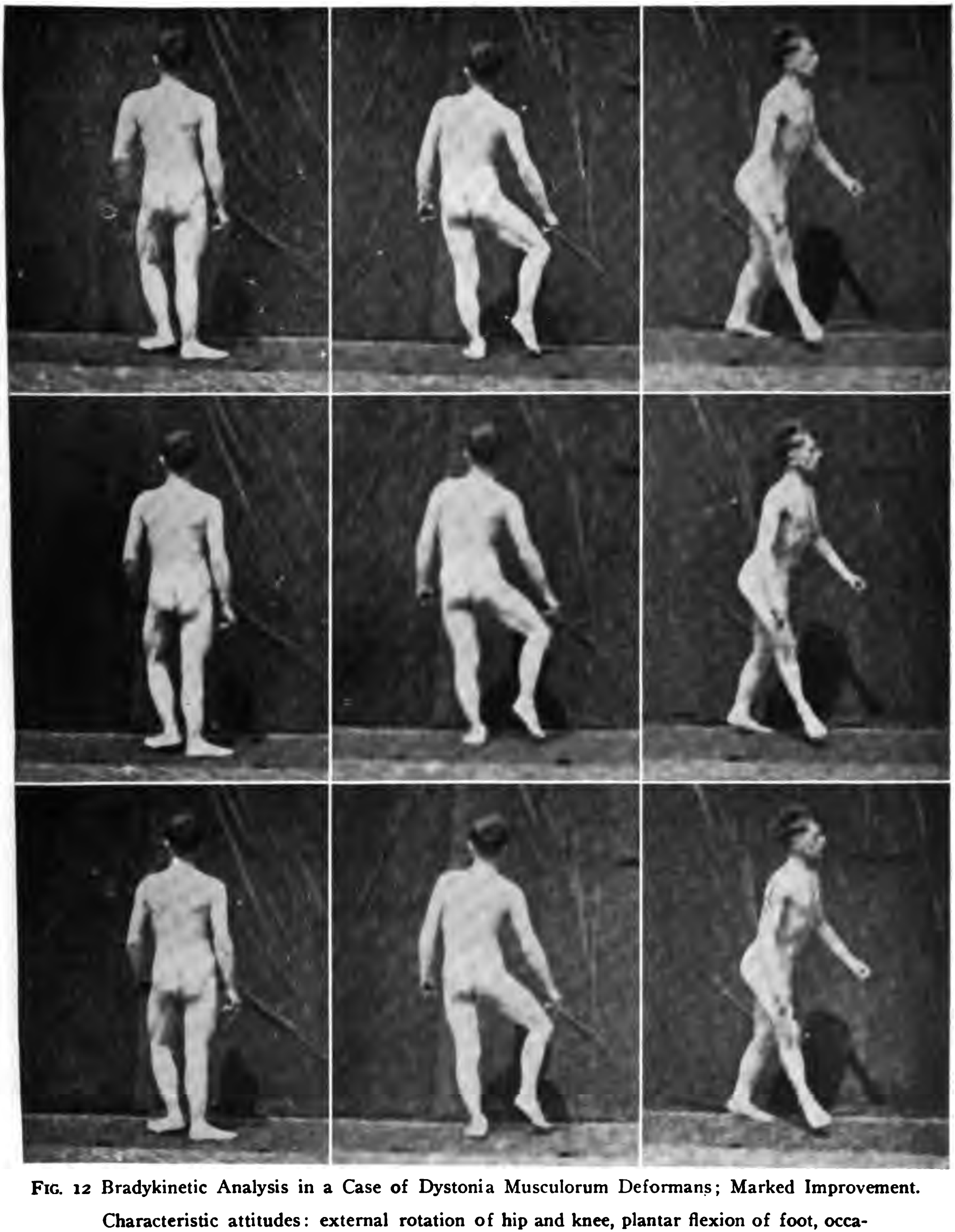

FIG. 12 Bradykinetic Analysis in a Case of Dystonia Musculorum Deformans; Marked Improvement.

Characteristic attitudes: external rotation of hip and knee, plantar flexion of foot, occasional eversion of foot.

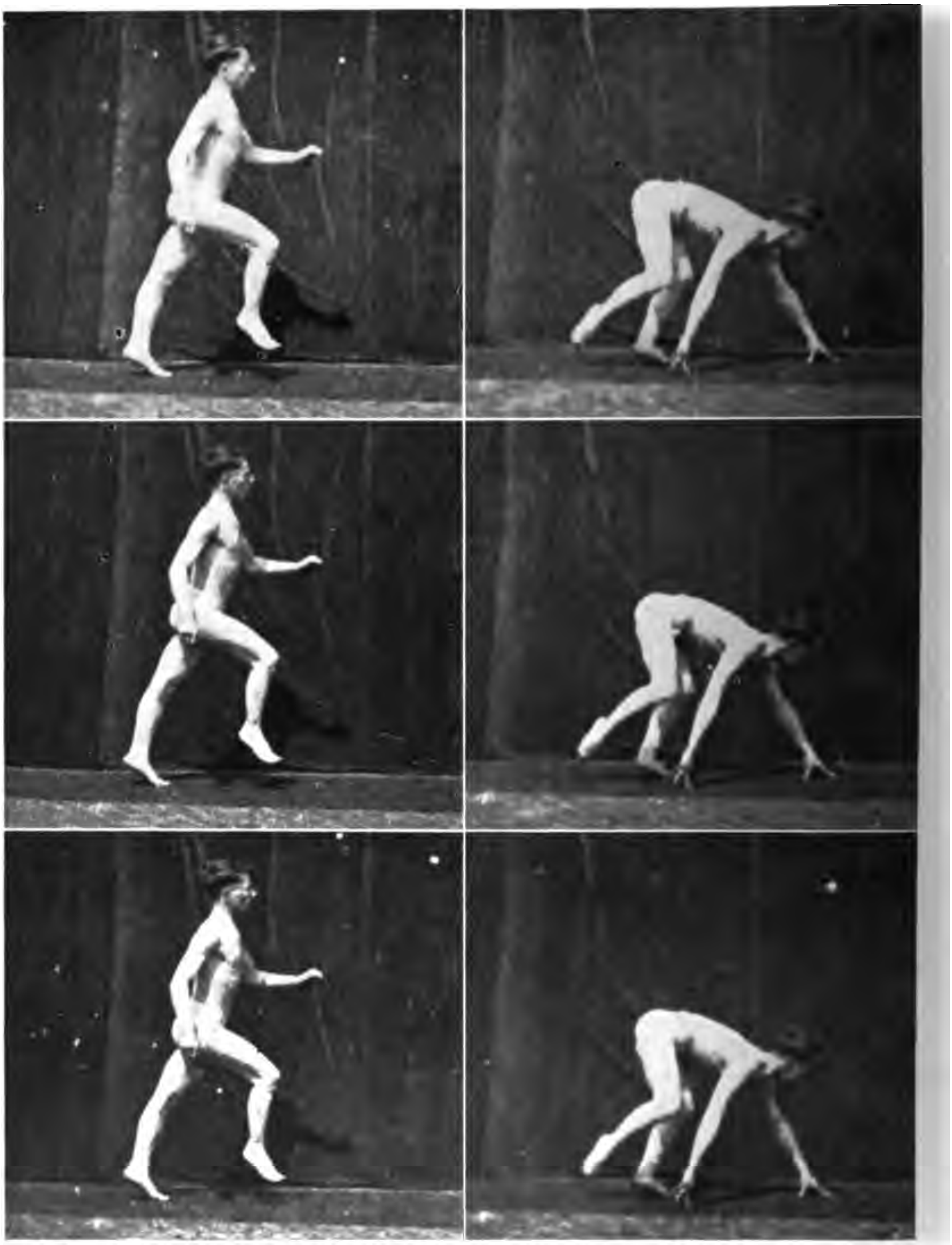

FIG. 13. Same as figure 12.

Showing marked improvement of muscle control and type of exercise helpful in re-education; demonstrating also as patient advances resting on fingers, ball of foot and toes, characteristic dystonia attitudes. This position on "all fours" as patient moves forward emphasizes the special deformity due evidently to defective postural fixation in the motor conflict which shoots, so to say, the pelvis and trunk upward in progression.

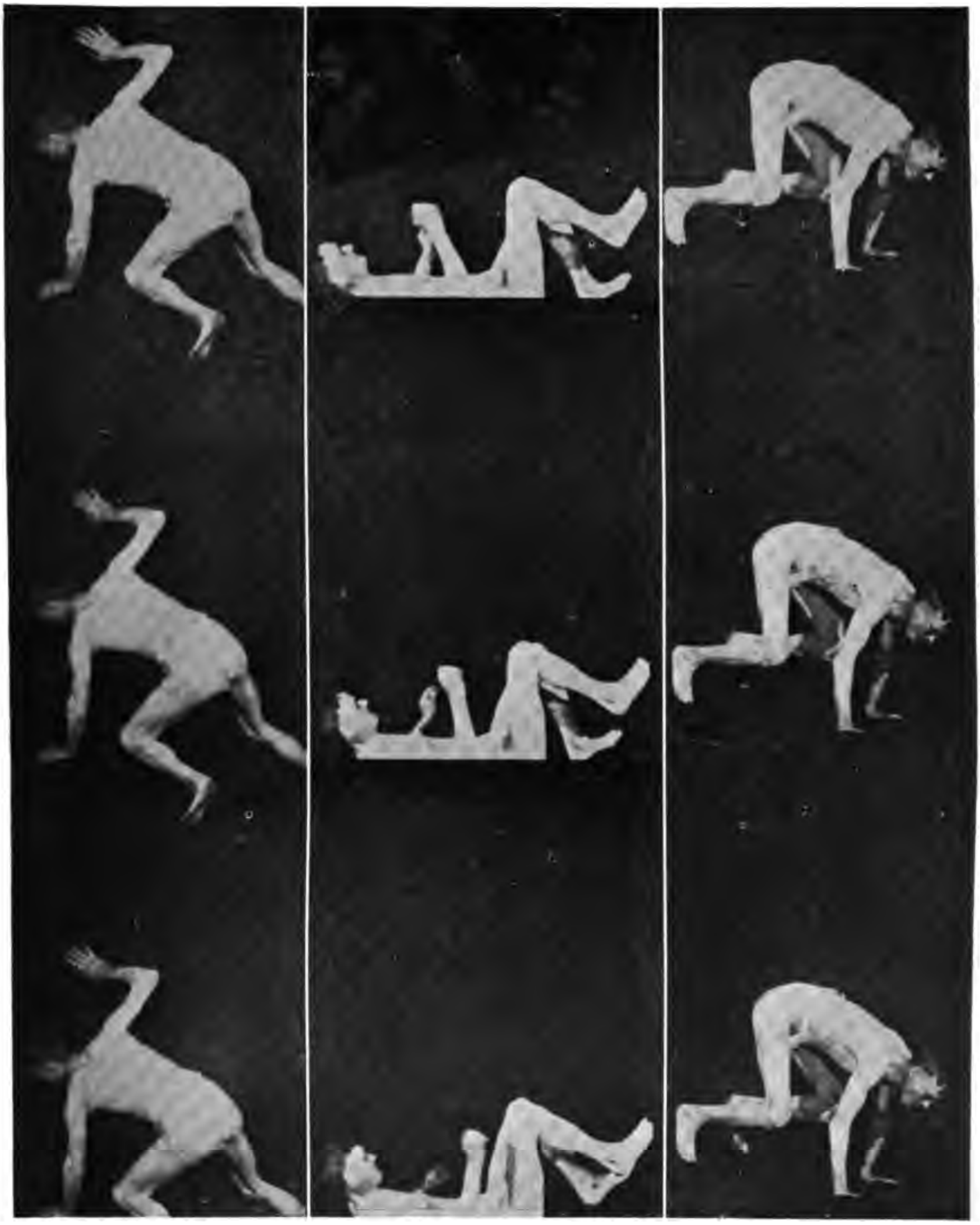

FIG. 14. Bradykinetic Analysis in a Case of Cerebellar Agenesis: Patient Falling and Rising.

As trunk approaches ground and efforts to balance are made, the normal interplay of muscle groups, agonists and antagonists, is seen to fail. Some group dissociation is seen upon efforts to rise. As patient attempts to gain upright position the defective integrative synergia causes vicarious efforts; the body is tossed upward and sustained for a moment upon the hands alone.

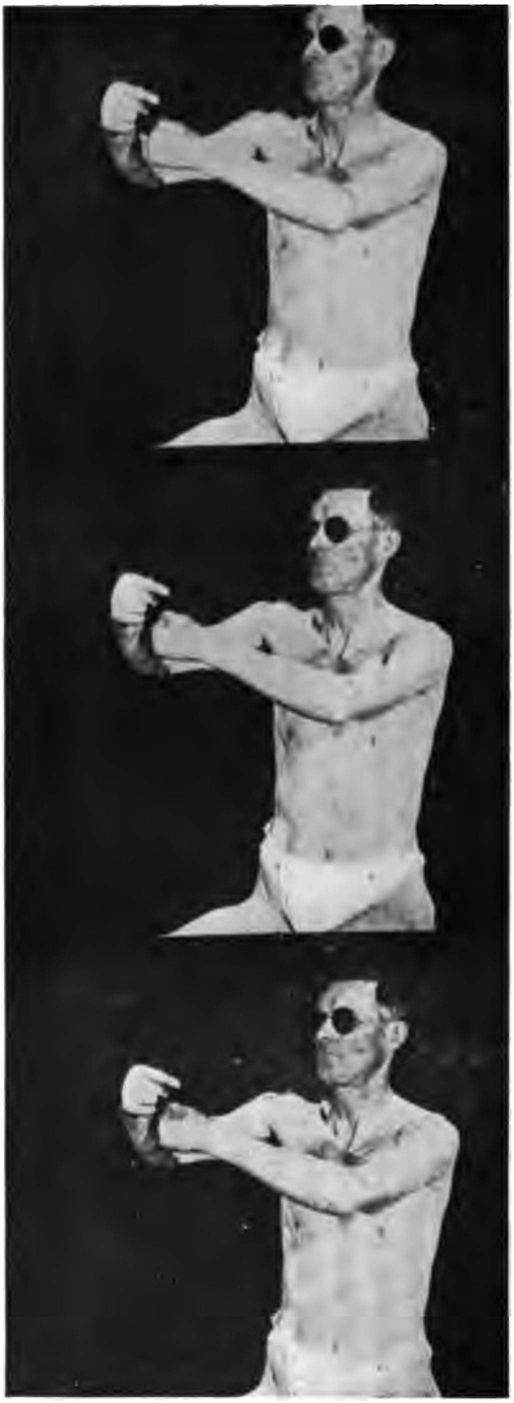

FIG. 15. Same Case as Figure 14; Showing Finger to Finger Test.

Careful observation shows the so-called tremor of the multiple sclerosis type to be the result of defective cerebellar function. The analysis by slow motion photography clearly defines the dissociation of the synergic unit. The serial pictures, of which the above illustrations are necessarily only fractional units, show the defective interplay of the agonist and antagonist and its corresponding check component thus mechanically defeating attainment of the purposeful movement.

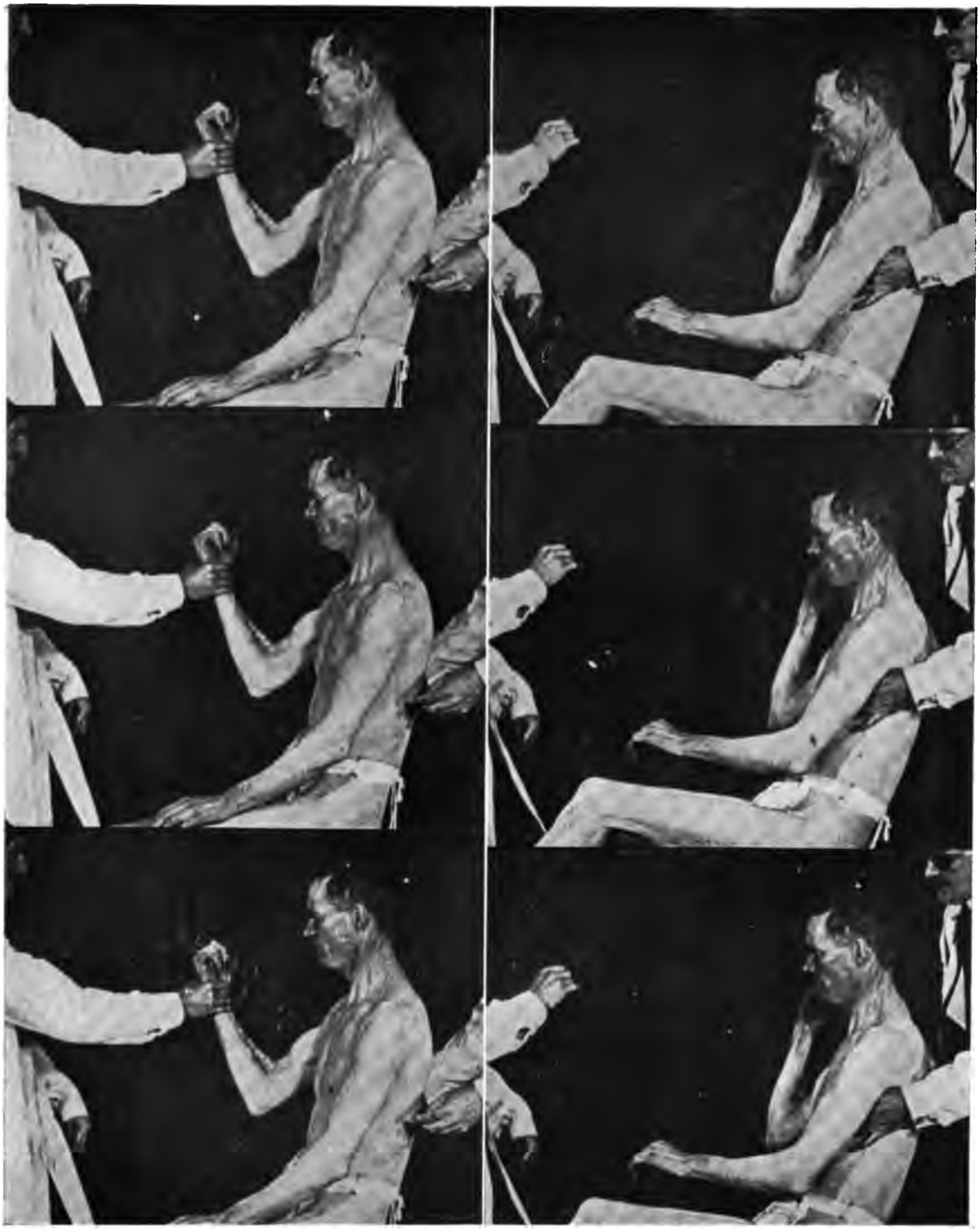

FIG. 16. Same Case as Figure 14: Showing the Abnormal Rebound Phenomenon of Gordon Holmes.

As the examiner grasps the patient's wrist and attempts to extend the flexed arm of the latter, the patient at the same time resisting, as the examiner suddenly releases the wrist, the patient's forearm rebounds forcibly, striking his cheek. This rebound, by analysis in slow motion, is seen to be due to failure of check component (the triceps) to promptly enter into coordinate function. Furthermore as indicating a general dissociation of synergic muscle group function, observe that all the segments of the body fail in associative activity resulting in general disequilibrium. (integrative dyssynergia).

SEGMENTAL OR NUCLEAR TRIGEMINAL SENSORY DISTURBANCE*

By I. S. WECHSLER, M.D.

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NEW YORK CITY

While the diagnosis of intrapontine lesions ordinarily does not present great difficulties, and is based mainly on alternating hemiplegias and crossed sensory disturbances, the question of distinguishing intra- from extrapontine disease occasionally arises. Attention has been called to the nuclear or segmental sensory disturbance of the trigeminal as a diagnostic point in the recognition of intrapontine lesions. Considering the comparative frequency of such lesions, the rarity of segmental sensory disturbance makes of this point a questionable diagnostic criterion. It has, in fact, been pointed out that intrapontine lesions may give radicular and even peripheral sensory disturbances in the distribution of the trigeminal.

It is not the object of this paper to enter into a discussion of the diagnostic differentiation between intra- and extrapontine lesions, but to report a case which showed what is known as the segmental or nuclear type of trigeminal sensory distribution. This type has been compared to "onion layers," that is, instead of extending across the face the divisions run outwardly from the nose to the ear. (Fig. 1.). Schlesinger and Déjerine have especially called attention to this fact. The following case illustrates such sensory distribution.

H. S., aged forty-five, male, tailor, married, was admitted to the Mt. Sinai Hospital on September 23, 1920. He complained of buzzing in the left ear and weakness of the left leg. His family history was negative. His wife gave birth to five children; she had no miscarriages. He had scarlet fever at twelve and is said to have had mild kidney disease for fifteen years. He denied venereal diseases. In 1902 he was operated on for an undescended testicle.

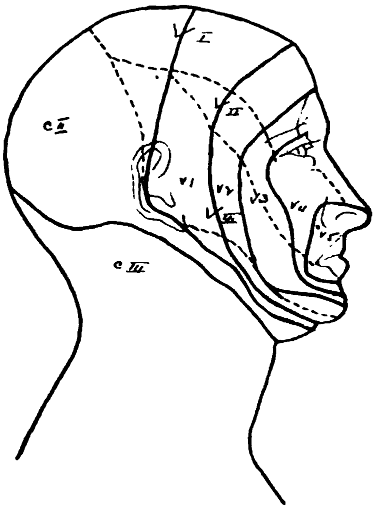

FIG. 1. Superimposition of peripheral and nuclear sensory trigeminal distribution. (After Déjerine.)

The present illness began gradually in June, 1919, with buzzing in the left ear. This was persistent, slowly became worse and led to impairment of hearing on that side. In September of that year a few herpetic vesicles appeared on the left side of the mouth, lasted a week and were followed by a pin and needle sensation on the left side of the face and forehead. At that time he had a shooting, intermittent pain which radiated over the entire left side of the face and forehead. This disappeared after a short while. About the same time he noticed a weakness of the left side of the face. In April, 1920, the left leg began to get weak, walking was impaired and he tended to fall to the left. In September, 1920, he again had an eruption on the corner of the left side of the mouth.

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*From the service of Dr. B. Sachs, Mount Sinai Hospital, New York, and presented at a Clinical Conference of the Neurological Department, Vanderbilt Clinic.

He complained of no vertigo, had no paralysis, no convulsions, no vomiting, no headache, no loss of consciousness, no tremors, diplopia or involuntaries. No fever at any time. His vision had become somewhat diminished. For the past few years he experienced difficulty in starting the stream in urination. He had polyuria, almost hourly, and nocturia four times. He had been impotent for the past few years. He noticed that he did not speak as well as he used to and that he could not whistle. He thought that taste had become impaired on the left side. He also noticed that he would cough on swallowing food.

PHYSICAL EXAMINATION

The gait was somewhat unsteady, but he did not stagger to either side. No Romberg. The finger to nose and heel to knee tests were normal, there was no pass pointing with either hand, but he showed left adiadochokinesis. His speech was a little thick. There were no tremors or other abnormal involuntary movements. The jaw-jerks were lively, the left being greater than the right. There was no clonus. The abdominals and cremasterics were absent. There was a definite right Babinski and a suspicious left. Motor power seemed generally somewhat diminished on the left side of the body. No atrophies.

Sensations. Pain, touch, temperature, vibration, position and stereognosis were normal all over the body and extremities. The right side of the face was normal. Beginning in the median line and extending outward in concentric fashion (Fig. 2) there was loss of touch, pain and temperature on the nose, about the forehead, eye, cheek and corner of the mouth, then diminution of these sensations on the face extending half way to the ear. Corneal sensation was lost on the left side. The left side of the tongue, the mucous membrane in the left nostril and left cheek also showed loss of sensation.

Cranials. Smell was normal. The fields of vision were contracted, more on the left temporal side. The fundi were normal. The pupils reacted promptly, were equal and regular. There were no ocular palsies. There was mild horizontal nystagmus, more marked to the right. There was weakness of the left facial, more pronounced in the lower branches. There was some question about impairment of taste on the left side. Bone and air conduction was diminished on the left side and somewhat impaired on the right. Weber lateralized to the right. Caloric stimulation of the left vertical canals gave no response, the left horizontal canals gave a faint reaction after four minutes. The right vertical and horizontal canals gave normal response in twenty-five seconds (Dr. Friesner). The palate and tongue were normal.

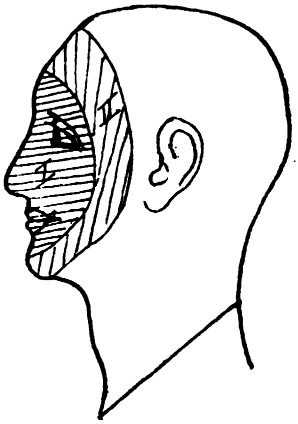

Fig. 2 Area I shows loss and Area II diminished sensibility.

The cranium was normal, not tender to percussion. The patient showed some emotional instability. Electrical reactions of the left face showed diminished galvanic and faradic response (Dr. Harris). A radiograph of the skull showed no signs of increased intracranial pressure, a large sella, definite in its outlines, with smooth floor and well-defined clinoid processes.

The general systemic examination was negative. The urine was normal; the blood and spinal fluid Wassermann reactions were negative.

DISCUSSION

While the sensory disturbance in the region of the trigeminus shown in this case seems to point to an intrapontine lesion, there are a number of facts both in the history and the physical findings which do not support the localization. If we accept the view that herpes is always due to a lesion of the sensory ganglion, the history of two herpetic attacks would point rather to extra- than intrapontine disease. The chronological order of the signs and symptoms as mentioned by the patient also speak in favor of an extrapontine, that is an angle lesion. The disease began with buzzing in the ear, later impairment of hearing, then disturbance of sensation in the face, later facial paresis, finally staggering and weakness of the left side of the body. These are almost classical symptoms of angle tumor, and the addition of choked disc would certainly have justified a craniotomy.

There are other facts which do not fit in with an intrapontine localization. The complete absence of sensory changes, aside from those of the face, are somewhat difficult to explain. That a lesion which affects the vestibular and cochlear divisions of the eighth, the sensory nucleus of the fifth, the seventh and the pyramidal tract (possibly both) should fail to compromise the adjoining fillet or the spinothalamics is rather unusual. So, too, one might expect dissociation between the cochlear and vestibular divisions of the eighth in intrapontine disease, which was not the case in our patient.

On the other hand, there are a number of factors against the diagnosis of extrapontine neoplasm. The condition was not sufficiently progressive; in fact, it had remained stationary for many months. Again, a lesion, such as neoplasm of fifteen or eighteen months' duration, in the posterior fossa and at the angle, should have given some degree of choked disc, some headache, and possibly vomiting. While, therefore, the localization is not by any means certain, the absence of choked disc (this is not infrequent in pontine neoplasms) and the segmental sensory involvement of the fifth are more or less substantial evidence of intrapontine involvement.

However, even without being able to commit oneself either as to diagnosis or localization the case is of unusual interest. It is reported especially for its clinical aspects and for the reason that the so-called nuclear or segmental sensory disturbance in the realm of the trigeminal is so rarely encountered.

[NOTE. For about one year after the patient was presented at the Conference and this paper was written his condition remained stationary. Then he developed choked disc and visual disturbance. This, in view of the history, definitely pointed to a neoplasm and justified a suboccipital craniotomy. A large, irremovable cerebellopontine angle tumor with a cyst overlying it was revealed at operation. The patient died the following day but no autopsy could be obtained.

The operative findings lend even greater interest to the case. In the first place it is possible for an angle tumor to go on for a very long time without giving signs of intracranial pressure. Secondly, it goes to show that when the history is fairly significant of an angle tumor, as that of this patient was, one is justified in making a diagnosis of intracranial neoplasm even in the absence of the so-called classical signs and symptoms such as headache, vomiting and choked disc. Thirdly, an operation would have been justified long before the onset of the choked disc and possibly would have saved the life of the patient. Fourthly, the history of herpes, the disturbances of hearing and the chronology of the symptoms should have made the diagnosis of extrapontine lesion definite, and we should not have allowed ourselves to be influenced too much by the sensory findings. Finally, it is quite probable that the neoplasm invaded the pons and accounted for the nuclear or segmental sensory disturbance of the trigeminal.]

CEREBRAL SUBARACHNOID HEMORRHAGE WITH RECOVERY

By CHARLES A. McKENDREE, M.D.

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Spontaneous rupture of cerebral vessels into the subarachnoid space has been recorded rather infrequently in comparison with the vascular accidents that involve the cerebral substance proper, the basal structures, and those in which the ventricles are flooded. Hence it may be of interest to present the following case which clinically represents this type.

As is frequently the case, the severe cerebral symptoms herein described have gradually cleared away and in a comparatively short time the patient has practically recovered.

L. A., aged thirty-one; American, married. Occupation, chauffeur.

Complaint. On October 3, 1921, he was admitted to the service of Dr. Frederick Tilney at the Neurological Institute, in a semiconscious state, suffering with excruciating frontal headache, and rigidity of the neck.

Present Illness. The patient was quite well up to the onset of his present illness. On September 11, 1921, he had a slight headache when he went to bed. During the night he dreamed he was confined in an elevator shaft into which the elevator was steadily descending. Terrified, he suddenly awoke with what he describes as a terriffic headache, extending all over the vertex and occipital areas. He immediately arose and walked about the room in an effort to relieve the pain. He had never experienced a similar condition. It seemed to him as though someone was "hammering inside his head." His wife reports that he fainted for a short time. Almost immediately he began vomiting, and this lasted in all about sixteen hours. About twenty-four hours after the onset he experienced a queer griping pain at the lower part of his spine, and he experienced "numb feelings" in the back of his legs when he walked. He does not recall having had any pain in his shoulders, arms or trunk at this time. He remained at home about six days, at which time he was sent into Roosevelt Hospital as a case of possible cerebro-spinal meningitis. When he was admitted, September 17, 1921, his chief symptoms were rigidity of the neck, and severe headache. In a few days these subsided, and when I saw him, he was not in great discomfort. Detailed examination at that time revealed absolutely nothing of a pathological nature except exhausting abdominal reflexes, a doubtful Kernig on the left, and a slightly rigid neck. The deep reflexes were equal and very active, but could not be considered pathological.

The patient was exceedingly difficult to puncture because of resistance and only one of the three attempts to obtain spinal fluid was successful. The specimen obtained was bloody, and did not coagulate. The culture was negative. The blood count revealed: hemoglobin 90 per cent; white cells 17,600; polynuclears, 85 per cent, and lymphocytes, 15 per cent. The blood Wassermann was 3 plus positive. The urine was practically negative, a very slight trace of albumen being detected on one occasion. The temperature reached 100° and 101° F. on two days but was at other times normal. The respiration ranged between 20 and 26, and the pulse rate between 80 and 96 during his stay in the hospital.

Although hemorrhage was suspected, we did not feel justified in placing much reliance upon the one cerebrospinal fluid specimen obtained, and the rapid subsidence of the presenting symptoms led me to diagnose the case tentatively as one of neurosyphilis, awaiting further spinal fluid data. On September 27, 1921, ten days after admission, he was discharged, practically free from symptoms.

Within the next week he visited us at the Vanderbilt Clinic on two occasions. On the second visit, Dr. Howe, at my request, examined him with great ease, and together we could find no abnormality. His only complaint at the time was rheumatic pains in the shoulders which lasted about three days. In the light of his experience he was ordered to have further blood and spinal fluid investigation and antispecific treatment.

On the night of October 2, 1921, he went to bed feeling well, and in the early morning was awakened with severe occipital pain. He arose, walked about, lighted a cigarette in an effort to relieve the pain but this was followed quickly by loss of consciousness, stiffness of his arms, legs and body (with no convulsive movements); his eyes were rolled upward and were glassy in appearance. He was cold to the touch and was very pale. Respiration was stertorous. There was no biting of the tongue nor bladder incontinence, but he was incontinent of feces.

The duration of the attack was approximately fifteen minutes. About ten minutes later a second occurred. This was followed by excruciating headache, and the patient appeared confused, singing at times, and saying "funny little things." At one time he complained to his wife of not seeing well with his right eye.

The next morning he was admitted to the Neurological Institute with the complaint mentioned.

Past History. The patient had typhoid and pneumonia as a child, an operation for right-sided hernia fifteen years ago, and influenza two years ago. He admitted repeated Neisserian infections, but denies lues by name and description. Apparently he never had any secondary signs—but he had been subject to so-called "quinsy sore throats" and had had one about four weeks before the onset of his illness.

He was married, and had one child living and well. His first wife died of tuberculosis. His present wife had had one or two induced abortions.

He is said to have had "tobacco heart" ten years ago. Bowels were regular and there was no difficulty in micturition.

As the patient was in such great distress upon admission, no detailed examination could be satisfactorily made. His pulse ranged between 40 and 50, and was irregular, but strong. The cranial nerves were negative, except for the findings of the ophthalmologist, Dr. Ward A. Holden, who records "the right vitreous filled with membranes of hemorrhagic origin—probably not recent; left normal." The motor and sensory systems were intact. Deep reflexes were all increased equally. There was a transient ankle clonus on the left. No Babinski or Hoffmann could be elicited. There was a bilateral Kernig and Brudzinski. The neck muscles were rigid and forced movements caused great pain.

Lumbar puncture performed at this time revealed very bloody fluid, which remained of the same character in two specimens. The patient slept a half hour immediately after the puncture because his pain was somewhat relieved, but it returned at the end of that time as severe as ever. In the course of the next three days he was punctured three times, and the fluid remained bloody, and did not coagulate. (The patient now has no recollection of having been punctured, but will not consent to further procedures.) He was continually moving about in bed, moaning, his hands constantly to his head, sleeping at intervals only when morphine was administered. He was noisy occasionally.

The blood Wassermann was returned 4 plus positive.

The blood counts made on five occasions are of considerable interest:

	Oct. 3rd	Oct. 4th	Oct. 5th	Oct. 7th	Oct. 24th
Hb.	80				
Color Index	.95				
R. B. C.	4,200,000				
W. B. C.	25,000	22,000	16,000	13,500	12,000
Neutrophils	93	81	75	70	08
Small Lymph	7	14	23	28	28
Large Lymph		3	2	1	1
Eosinophils					1
Large Mononuclear		2			1

The spinal fluid showed a very heavy blood sediment, bright red in color, with no coagulation, 2500 cells. The Wassermann reaction was, of course, strongly positive.

The urine was negative throughout.

The temperature reached 101.5° F. the first three days in the hospital; then for three days, 100.5° F., and normal since. Respirations were never below 20, the highest rate being recorded as 24. There was no Cheyne-Stokes type at any time.

The pulse rate was below 65 for two days, but quickly came back to an average of 80.

The headache gradually subsided, although a steady dull headache, chiefly frontal, persisted up to recently. Five days after admission he was mentally clear, and has remained so.

On October 10, 1921, a left facial weakness of the central type was noted, but it was not marked. This has persisted to the present time.

Dr. Holden reported no change in the ocular apparatus on October 13th, but six days later there were evidences of fresh hemorrhage. The red reflex could not now be seen in the right eye.

The stiff neck has entirely disappeared, and the patient, except for poor vision of the right eye and general weakness, feels perfectly well.

NEUROLOGICAL EXAMINATION

On October 16, 1921, it was possible to examine the patient thoroughly without subjecting him to great discomfort.

Abnormal Attitudes and Deformities. None.

Gait. Normal in all phases in walking.

Coordination. There is very slight swaying with the eyes closed; none with eyes open. Patient stands on the left foot better than on the right, but does fairly well. Finger-to-nose, and finger-to-finger tests are normally performed. Heel-to-knee and along shin tests are well done on both sides, with very slight unsteadiness. Diadochokinesis is normal.

Skilled Acts. Speech is normal. Test phrases are accurately repeated. There is no aphasia.

Abnormal Involuntary Movements. None can be observed.

Reflexes. All the deep reflexes of the upper and lower extremities are lively but equal. There is no ankle clonus. The abdominal, epigastric and cremasteric reflexes are active and equal. Plantar flexion is normal. No Babinski or modifications are present.

Muscle Strength. There is no noticeable loss of power in any of the muscle groups.

Muscle Status. Volume and contour are normal. Tone is normal, and consistency is rather soft.

Sensation. Touch (acuity), pain, vibratory and muscle tendon sensibility are normal throughout. Stereognosis is normal.

Cranial Nerves. The fundi are reported above. The pupils are equal, regular, central, reacting to light and convergence. There is no strabismus but a moderate exophoria on the right. Nystagmus, ptosis and exophthalmus are absent. Hearing is normal; Weber and Rinné tests are properly responded to. Motor and sensory fifth are normal. There is apparently slight weakness of the left seventh (lower two-thirds) upon inspection, but in the tests it completely disappears. The ninth, tenth and eleventh nerves show no abnormality. The tongue protrudes in the midline.

Mental Status. Although the patient has no recollection of his admission to the hospital and the first three days' experiences there, he is now entirely clear, has perfect orientation, and is most cooperative.

Systemic Examination. The tegumentary and glandular status is normal. The heart is negative, blood pressure 110. The respiratory, gastrointestinal and genito-urinary systems are negative.

SUMMARY OF ESSENTIAL OBSERVATIONS

1. Sudden onset of peculiarly severe headache, followed rapidly by vomiting.
2. Rigidity of the neck, and pain in the shoulders and lower back, which gradually subsided.

3. Three weeks later, sudden recrudescence of headache, vomiting, stiff neck, convulsions of the purely tonic type, delirium, etc.

4. Bradycardia, persisting leucocytosis, strongly positive Wassermann.

5. Intra-ocular hemorrhages on the right.

6. Left facial paresis, and transitory clonus.

7. Bloody spinal fluid on four occasions.

8. Gradual and progressive recovery.

In the study of the sequence of symptoms in this case, it is quite evident that the patient had two distinct vascular accidents, the second being much more extensive than the first. The underlying pathological factor being syphilis, without reasonable doubt, it is most probable that the hemorrhage resulted from the rupture of a very small aneurism. As the patient suffered such a small amount of hemorrhage, however diffuse, I believe it is most probable that the ventricles and the cisterns have not been invaded. Certainly he presented none of the characteristic signs and symptoms described a few years ago in the *Neurological Bulletin* by Tilney and Casamajor, and the patient's recovery makes such a serious involvement quite unlikely.

The eye symptoms can be explained only upon the basis of a simultaneous rupture of an intra-ocular vessel during the convulsions.

The exact site of the origin of hemorrhage must be problematical in this case as there have been so few localizing signs. However, bearing in mind the transitory clonus of the left ankle, and the slight facial weakness on the left, which is present to-day, I venture to suggest that the small aneurism was located on the pia-arachnoid on the right side, possibly on the convexity of the brain.

The prognosis in this case should be good, as antispecific medication, thoroughly and continuously employed, is especially favorable in this type of vascular lesion.

DISCUSSION

Dr. Wechsler. I agree with Dr. McKendree that the case is one of luetic aneurysm, and in this connection I want to report the case of a young boy who while driving a bicycle fell off and became unconscious. I do not know whether he fell as a result of the unconsciousness or became unconscious after the fall. He was taken to Bellevue Hospital and developed ptosis of the right lid. Spinal puncture showed bloody fluid. He was unconscious for several hours, had complete paralysis of the third nerve. He recovered from that after a few days and had a similar experience several weeks later. He became unconscious again and had bloody spinal fluid. From this, too, he recovered, but the original paralysis had remained. That he did not have syphilis was shown by the negative Wassermann in blood and spinal fluid. The tentative diagnosis that was made was encephalitis.

Dr. Cornwall. Subarachnoid hemorrhage is more frequently seen in cases of miliary aneurysm of the basilar or one of the communicating arteries. These may or may not be due to lues. In this case I see no reason why the location might not be where Dr. McKendree has put it.

The question of blood in the spinal fluid is an important one. I am perfectly confident that I have seen a spinal fluid remain bloody for several days after accidental injury to the vessels at the point of the puncture.

I think that Dr. McKendree's diagnosis is the correct one.

Dr. Casamajor. Personally, I believe that this was a real hemorrhage into the arachnoid; but how one is going to tell whether the bloody fluid in these cases is due to hemorrhage or to puncture of a small vein, I do not feel as though I were in a position to say. The relation to syphilis here I think is very clear. A miliary aneurysm might easily be on a syphilitic basis. One should not be surprised that syphilis would cause hemorrhage; it is surprising that it does not cause it more often. Endarteritis may go on to the formation of aneurysm in some vessel, and then thrombosis increasing the pressure ahead would give most beautiful opportunity for the blowout of the aneurysm. The vessel that would blow out here is much more apt to be an arachnoid than a pial vessel. The arachnoid vessels are small. The pial vessels lie close to the brain surface, so that a hemorrhage from an artery would be much more apt to infiltrate through the pia with the pial spaces ballooning up. That would give a greater spread of hemorrhage which is what we saw in a case Dr. Tilney and I described.

I think the dural vessels can be excluded because the dural vessels, especially in the skull, are bone vessels and lie on the outer surface of the dura; and one would have difficulty in conceiving of a hemorrhage penetrating the dura.

As to the question of leucocytosis, while we do get it, usually it is not present in such a small hemorrhage as this.

The interesting thing is that this man should have had two arachnoid hemorrhages and survived so well, which I think would have been impossible were the hemorrhage in the pia.

DR. MCKENDREE. With reference to the question of the value of bloody spinal fluid, I would say that I believe that two or more specimens containing blood should always receive serious attention. In this particular instance, I know that had I not been so conservative in accepting the findings at first, the second and more serious attack which the patient suffered would have been averted. While at the Institute, the three punctures were performed very easily, the bloody fluid ran steadily and did not coagulate. They were done at intervals of twenty-four hours or more, and with the least possible trauma. Four specimens of like character, together with the leucocytic reaction, seemed to me to justify without question the diagnosis of subarachnoid hemorrhage.

Moreover, the symptoms presented—headache, pain in the shoulders, pain in the lower spine, rigidity of the neck, Kernig, etc., together with the laboratory findings—clearly indicate the nature of the cerebrospinal symptoms.

I would like especially to emphasize the point that in this case, the convulsive movements were of the tonic type and at no time presented the adduction and abduction movements of the upper extremities such as were so ably described by Drs. Tilney and Casamajor in their case of ventricular and cistern hemorrhage.

In Dr. Wechsler's case, it is quite possible that the hemorrhage was more or less circumscribed and encysted. In the case presented today it is apparent that the hemorrhage must have been comparatively small but diffuse, and resulting in very little compression.

I believe that we do not encounter repeated ruptures of this type, because the nature of the lesion is usually discovered, and subsequent treatment, especially in those cases in which syphilis is the underlying factor, is successful in averting further accidents.

THE SIGNIFICANCE OF NYSTAGMUS IN A LESION OF THE ABDUCENS NUCLEUS

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In spite of the excellent work done by a large number of reliable investigators, the pathways of the cerebello-vestibular mechanism remain ill-defined. The physiological significance of the three or four morphologically different cell groups in the vestibular area of the medulla oblongata is quite unknown. The connections between the vestibular and the oculogyric nuclei, maintained on a firm clinical basis, are from an anatomical standpoint involved in greater obscurity today than they were twenty years ago. In 1901 Frazer conducted an investigation into the relations of the posterior longitudinal bundle and the vestibular nucleus. He concluded that "very many fibers pass from the posterior longitudinal bundle to the third and fourth nuclei, some of these being derived from the sixth nucleus, but by far the larger number from Deiters' nucleus."

This conclusion was based on a tracing of degenerated fibers after *complete* destruction of Deiters' nucleus in monkeys. The very completeness with which any special part is destroyed in the complex and closely packed region of the cerebello-medullary junction, must make the tracing of degenerated fibers from that special part a task fraught with a certain degree of uncertainty. Van Gehuchten, however, in a similar research, agrees with Frazer's findings. He found degenerated fibers going from Deiters' nucleus to the same and to the opposite side of the posterior longitudinal bundle to reach the nuclei of the third and fourth nerves. He corroborates emphatically the findings of Probst, whom he quotes as follows: "I could show with certainty fibers going from Deiters' nucleus mainly to the same side, and in smaller numbers to the opposite side of the posterior longitudinal bundle. In the posterior longitudinal bundle the fibers . . . give off very rich numbers of collaterals to the nuclei of abducens, trochlearis and oculomotorius. These fibers then connect the nucleus of Deiters . . . directly with the nuclei of the ocular muscles. . . "

Lewandowsky is similarly unequivocal respecting the existence of vestibulo-oculogyric connections. Cajal, in his studies of silver-impregnated sections of these parts, complained of the extreme fineness of the fibers connecting the vestibular and the oculogyric nuclei. He concludes, however, that "the existence of vestibular fibers in the posterior longitudinal bundle . . . agrees with the findings of physiologists of a mechanism for compensating associated movements of the eyes when the head and body change their balance." The statement is pregnant with a germ of uncertainty and agrees with the drawings of the sections on which Cajal based this conclusion. It must be confessed that these drawings are not entirely convincing, it is not possible to see in them the connecting pathway in question with any high degree of certainty. The admirable effort of Sachs and Alvis at unravelling the vestibular mystery has thrown a good deal of doubt in the pathway connecting Deiters' with the oculogyric nuclei. One of the chief merits of their work is the convincing manner with which they have pointed out a misleading factor in all such experimental investigations—the extreme difficulty of producing a lesion of the parts in question without at the same time injuring neighboring structures. In this connection their conclusion, which is based on a more extensive and more exhaustive study than had been heretofore undertaken, is that "the fibers running from Deiters' nucleus anteriorly in the posterior longitudinal bundle are few in number and stop before they get to the third and fourth nuclei so that it is doubtful whether there is any connection between Deiters' nucleus and the other nuclei . . ." Another conclusion at which these investigators have arrived, and which involves the subject in still greater confusion, is that "all anatomic evidence to support Bárány's contention as to the connections between Deiters' nucleus and the cerebellum, is lacking." They found, however, that some of the fibers of the vestibular nerve end in the nucleus tecti of the vermis—a feature which compensates considerably for the confusion introduced.

The following case brings home the need of further anatomical and physiological work in this direction.

Michael F., an Irishman, aged fifty-two, a porter by trade, came to the Neurological Division of the Vanderbilt Clinic on July 18, 1921, with the following complaint:

On June 1, 1921, he arose in the morning to find that his face was drawn to the right side and that he could not close his left eye. On his way to work the same morning, the street in the distance appeared double. The condition grew rapidly worse in the course of that day and then remained stationary until his appearance at the clinic.

Personal History. He had been a very healthy man all his life, a hard and steady worker. He had never suffered from any serious illness nor sustained any injury. He was rather an excessive smoker. His consumption of alcoholic beverages was never to the extent of causing intoxication, but he had drunk "reg'lar," as he expressed it. This regular drinking consisted in the average daily consumption of about a pint of whiskey and a gallon of beer for a period of some thirty years before prohibition, and of about one-third to one-half of those quantities since. He had been married for the last eighteen years. His wife had had no miscarriages and had given birth to four children, three of whom died from diseases incident to childhood. His present trouble came on overnight, without any preceding illness. Outside of the annoyances incident to this particular trouble, he suffered no physical discomfort of any kind.

Physical Examination. This was done some six weeks after the onset of the trouble and showed as follows:

The patient was a strong, healthy-looking workingman. His weight was 187 pounds. His face was markedly flattened on the left side, the mouth and nose were drawn to the right. His left palpebral fissure was much larger than the right. He had an internal strabismus of the left eye.

The *gait* was normal in all its phases. He could walk on his toes and heels, backwards and sidewise and, with a little difficulty, along a straight line.

The *associated movements* of the trunk, arms and head were those of a healthy person.

Coordination. He could stand with his feet close together, and with his eyes either open or closed, with the ordinary amount of slight swaying. The finger-to-nose and finger-to-finger tests were carried out well. There was perhaps a slight deviation to the left in pass-pointing with both hands, but outside of this there was no dysmetria.

Diadochokinesis was normal.

There was no defect in his speech.

The *reflexes*, both deep and superficial, were normal and equal on both sides.

The muscular strength of his limbs, body and neck was good.

There was no impairment of any of the sensations.

Cranial Nerves. The external rectus muscle of the left eye was paralyzed. The patient could not turn the left eyeball outward, that is, to the left of the midline.

The right internal rectus was weak in an attempt to move the right eyeball to the left in conjunction with a similar effort in the same direction on the part of the left eye. The same movement in convergence, however, was normal.

When the patient turned his eyes in a horizontal direction to the right, there was a pronounced rhythmical nystagmus of an amplitude of nearly a millimeter, repeated at the rate of about four per second. The movement continued as long as the eyes were turned to the extreme right.

The pupils were of normal size and shape, and they reacted well to light and to accommodation. A rough test showed no narrowing of the fields of vision.

The facial muscles of the left side were paralyzed and flaccid. He could not whistle, close the left eyelids nor wrinkle that side of his forehead.

There was a definite loss of the gustatory sense on the anterior two-thirds of the left side of the tongue.

There was no hypo- nor hyperacusia on either side.

There was nothing abnormal about any of the other cranial nerves.

The patient's mental status was good. There was apparently nothing wrong with his internal organs. His blood-pressure was 140 / 90. The blood Wassermann test was negative.

Since his first appearance at the clinic the patient has been observed a number of times. There has been a progressive improvement of the symptoms in the following order: The gustatory sense returned by August 29th. The facial paralysis was next to improve; by September 12th the deformity was no longer conspicuous. Two weeks later the face appeared almost normal: the patient could whistle, wrinkle his forehead and close his left eyelids. The nystagmus showed a progressive diminution of rate and total duration at the same time that its amplitude became rather wider. The paretic condition of the right internal rectus muscle upon an effort at conjugate movement of the eyes to the left was considerably improved. At the time of this writing the patient shows no easily noticeable abnormality of the cranial nerves except one—a paralysis of the external rectus muscle of the left eye.

Localization. Since all the symptoms in this case appeared suddenly and simultaneously, without any preceding illness, one is justified in thinking *a priori* of a single lesion in the region of the left abducens nucleus. Among the structures that may possibly be involved in such a lesion the anatomical relations of some are well established, while the relations of others are still largely wanting in anatomical demonstration. What is true of the anatomy of the parts involved is true as well of their physiology. The latter is defective in point of correlating certain clinical manifestations of disease of those structures with their alleged normal functions.

The fibers of the facial nerve form a loop around the nucleus of the sixth nerve; that much is known with certainty, and may be seen in any well-stained section of that level. A lesion extending slightly beyond the margin of the abducens nucleus must implicate the loop of the seventh nerve.

The nervus intermedius, upon entering the pontomedullary angle between the seventh and eighth nerves, proceeds mesially for some distance, then bends downward to reach, probably, the nucleus of the fasciculus solitarius. At its bend it is situated within a short distance of the nucleus of the sixth nerve and is apt to be involved in a lesion of that nucleus, extending downward and outward for a few millimeters.

It goes without saying that a complete lesion of any nucleus must cut off the pathways connecting it with all other structures. On this basis we may account for the inability of the right internal rectus muscle to turn the eyeball to the left in an effort at conjugate movement of both eyes, while it is still able to turn the eyeball to the left in a movement of convergence.

The structures responsible for the nystagmus are much more difficult to locate in our case. It is not a *paretic nystagmus*. The left external rectus muscle is paralyzed completely by a complete destruction of the left abducens nucleus. The right internal rectus muscle functions normally in all but a conjugate movement to the left. There is no question about the normality of the muscles which turn the right eyeball to the right; and even the overaction of the left internal rectus does not explain the permanent oscillation of both eyeballs in a horizontal movement to the right. A single quotation from Wilson and Pike's account of their experimental work in this connection will suffice to indicate the difficulty. "Nystagmus occurring in the dog and cat," say these authors, "as a result of labyrinthine stimulation or destruction, consists of two phases, a slow deviation followed by a quick return and varying definitely in direction with the labyrinth which is stimulated or destroyed. . . . . The slow deviation is not affected by removal of the cerebellum, nor is it affected by complete removal of the cerebrum, optic thalamus, partial removal of the posterior corpora quadrigemina and longitudinal splitting of the cerebellum. In short, if the anatomical path from the labyrinth through the vestibular nuclei to the eye nuclei (posterior longitudinal bundle) be intact, the deviation can be produced by stimulation, and appears after destruction, so soon as the animal begins to pass out from under the influence of the narcotic."

The striking statement in the foregoing quotation is that which refers to the same effect produced by two opposite causes—an oscillation of the eyes resulting alike either from stimulation or destruction of the labyrinth. The puzzle is susceptible of but one solution—that stimulation or destruction of the labyrinth alike bring about a disturbance in some organ whose function it is to coordinate the working of the oculogyric muscles. We know, moreover, that the nystagmus resulting from the destruction of the labyrinth is of temporary duration; and this, too, can be explained only on the supposition that the disturbance brought about in this manner in the organ endowed with the function of coordination was only temporary. Sachs and Alvis' findings give a good deal of strength to the foregoing supposition. If fibers of the eighth nerve enter the nucleus tecti of the vermis, we may suspect that an impulse propagated along that nerve would cause some disturbance in the normal functioning of the cerebellum; and that the impulse thus propagated being of a temporary character, the disturbance caused by it would be temporary. We can understand, furthermore, why the removal of the cerebellum would not influence the nystagmus brought about by the destruction of stimulation of the labyrinth. Since nystagmus is not a function but a disturbance of function—the function of muscular coordination—there is no reason why that disturbance should abate with the removal of the organ which presides over muscular coordination.

That the nystagmus which ensues upon destruction or stimulation of the labyrinth is due not to the effects of such destruction or stimulation upon the vestibular nuclei but to a disturbance in cerebellar functioning, appears rather reasonable from the following purely theoretical consideration:

It appears to be the general agreement that the function of the cerebellum is to coordinate the contractions of muscles and groups of muscles in the various positions assumed by the body and limbs; and that the vestibular apparatus is an adjunct to the cerebellum, endowed with the special function of receiving and transmitting impulses in response to the stimulus of gravitation. A movement of any part of the body which results in a change of position of the center of gravity calls for the harmonious working of two mechanisms: one, a mechanism for coordinating the contractions of certain muscles and groups of muscles; the other, a mechanism for furnishing correct information from moment to moment respecting the effect of the particular movement upon the position of the center of gravity.

Reasonable as it is to suppose that the vestibular mechanism is called upon by the cerebellum to furnish information respecting the position of the center of gravity of every movable part of the limbs, the body and the neck, it is not entirely reasonable in the case of the eyeballs. The position of the center of gravity is not changed by the rotation of a spherical body, of homogeneous consistency, around any of its axes. The eyeballs, in this respect, are practically spherical. In handling a dead animal its movable parts change their relation to each other with every change in their centers of gravity; while its eyeballs retain the same relative positions to the orbits throughout. The centers of gravity of the eyeballs remaining unaffected by the changing positions of the body, it is difficult to see what the vestibular apparatus has to do with the eyes. The business of adjusting the visual axes to the movements of the body and head would, therefore, be entirely between the optic mechanism and the cerebellum.

Since the exact anatomical pathways of the vestibular area are in doubt, we are thrown upon the expedient of weighing probabilities. The probabilities are strongly in favor of pathways from the vestibular nuclei in the medulla to all the motor nuclei in the central nervous system, with the exception of the nuclei of the oculogyric mechanism. For the proper coordination of this mechanism the cerebellum does not stand in need of information respecting the center of gravity of the eyeballs; that factor remaining constant, the cerebellum can accomplish the mechanical part of the task with the help of the optic apparatus only.

If this line of reasoning be correct, and the nystagmus in the case of our patient be a mode of incoordination of the nature of dysmetria, we must postulate in his particular case an injured conduction pathway from the cerebellum to the nucleus of the sixth nerve, an interrupted rubro-abducental tract.

Little need be said about the nature of the lesion because little can be said. The suddenness of the onset of the symptoms and the apparently small extent of the lesion speak for a thrombus occluding a small artery, probably one of the radicular branches.

The diagnosis of a single small lesion involving the left abducens nucleus and extending in a narrow rim around it, is to a certain extent supported by the subsequent developments. In the recovery from such a lesion, it might be expected that the symptoms produced by the injury of the parts about the periphery of the nucleus would pass away first. This has happened in the case of our patient. The symptoms produced by the involvement of the structures surrounding the abducens nucleus passed away in the order of their distance from it. The nervus intermedius was first to emerge; it was followed by the seventh nerve; the conjugate pathway to the nucleus of the opposite internal rectus muscle, as well as the pathway which carries the impulses of muscular coordination, recovered last; while the most central part of the lesion has left the nucleus of the abducens nerve permanently injured.

DISCUSSION

Dr. Sands. It seems to me that this case is one of a group that have come to this clinic of late with, apparently, disturbance referable to the extra-ocular mechanism without any other symptoms such as headache or fever or more serious disturbance.

A pathological basis such as a hemorrhage might account for the lesion which has now become absorbed. I do not think, however, that a hemorrhage in that area would give such a good recovery. Encephalitis seems a more reasonable diagnosis.

In spite of this, I am not sure whether the man has not a luetic lesion attacking the sheaths of the nerves rather than the nuclei themselves. I have never seen a real nuclear sixth lesion; I do not think they are very common. When you think of the path the sixth nerve has to travel, it would seem that any meningeal condition there would certainly strike it.

Dr. Strong. As to the location of the lesion, those who advocate an external, say a meningeal, lesion, overlook the presence of a deviation of gaze, i. e., a paresis of congugate eye movements. The presence of this phenomenon compels a central localization of the lesion.

Regarding the point raised about taste being affected, the location where the pars intermedia fibers terminate or bend caudad is not far from the abducens nucleus, being somewhat caudally as well as laterally located with reference to the latter.

The remarks made concerning vestibular nystagmus appear to ignore the fact that cerebellar nystagmus differs in several respects from the former.

Dr. Osnato. It does not seem to me that we ought to indulge in this sort of anatomical speculation until everything has been done from the clinical standpoint to clear up the case. There is no reason why any case of suspected vestibular origin should not be subjected to complete Bárány investigation. I have examined this man's nystagmus and I am not any too sure that it was of the type that we clinically recognize as either vestibular or cerebellar. There are three kinds of nystagmus: One is due to paretic imbalance, which is very common. There is no telling whether the man has had this type of nystagmus before this condition commenced. Then there is the vestibular nystagmus, and the cerebellar type that we see most typically in multiple sclerosis. That is unceasing, even being present on fixation, and is usually rotary and wild. This man's nystagmus disappears on fixation. It is only brought out on looking to the right; it is horizontal; it has not a rhythmical slow and quick jerk. It looks very much to me like a paretic ocular nystagmus due simply to imbalance in the muscles, entirely peripheral in nature. I do not think it has been at all proven that this is a vestibular or a cerebellar nystagmus. I do not think we should go any further with the anatomical explanations that have been advanced until at least this point has been decided in each case presenting the symptom of nystagmus.

Dr. Wechsler. Pathologically this case may be a luetic one; clinically it falls into the group comparable to posterior cerebellar artery thromboses. I should think that this man was entitled to anti-luetic treatment in spite of the fact that he had a negative Wassermann.

Dr. Cornwall. I think this man is old enough to have a thrombosis from arteriosclerosis and it should not be considered necessarily luetic without some proof. I do not believe that the underlying pathology is from a hemorrhage, but rather from thrombosis with resultant softening.

Dr. Casamajor. This was a thing of sudden onset and various possibilities have been suggested. The question of encephalitis I think is rather doubtful. It would be a small kind of encephalitis to pick out the position of this lesion. The question of hemorrhage can also be excluded. Hemorrhage in the soft tissue of the gray matter infiltrates pretty easily and would involve more than is seen in this case. Dr. Cornwall has brought up the point of thrombosis. There is a fairly large artery passing in with the seventh nerve to the sixth nerve nucleus. The sixth nerve nucleus, apparently, has a good many capillaries. I should imagine that a thrombosis might account for the sixth nerve paralysis without necessarily involving permanently the fibers of the seventh.

The question of localization is another matter. While one does not like to think of a small lesion in such a structure as the sixth nerve nucleus, yet I think the burden of proof is on the person who does not accept that lesion. It would explain the involvement of the motor seventh. Nor do I think that the point that the pars intermedia is also affected is really well taken in this case, because it was only partly affected as it easily could be in such a lesion. I think a sheath lesion is also out of the question. One could not think of a lesion affecting the seventh nerve without also affecting the eighth and that is not the case. A lesion large enough to affect both the sixth and seventh nerves would strike much more of the cerebellar mechanism and give symptoms more like cerebello-pontine lesions.

The anatomy of the nystagmus I do not think is a question we can discuss too much. We have become too prone to accept localizations, not only in nystagmus but also in regard to coordinative eye movements, that are physiological and not anatomical. This question is still open and Dr. Rosett has offered us some explanation; but I feel that the nystagmus in this case has been much better explained by Dr. Osnato's suggestion of a paretic nystagmus.

Nystagmus lasts a long or a short time according to the severity of the paretic lesion. And much depends on the individual who has it. There is an attempt of the individual to overcome the resulting diplopia which makes the nystagmus. This sort of thing will continue until the individual psychically cuts out one image and reverts to monocular vision. That, I think, is rather a different thing from the real nystagmus due to difficulty with the vestibular mechanism. Dr. Osnato spoke of the nystagmus in multiple sclerosis being cerebellar nystagmus. I am not in sympathy with that view. The nystagmus in cerebellar lesions that have come in my experience has been of the vestibular type. The multiple sclerosis nystagmus is much more nearly akin to the fine tremors of the late multiple sclerosis than to cerebellar ataxic movements.

ARTHRITIC MUSCULAR ATROPHY WITH REPORT OF A CASE SIMULATING ANTERIOR CRURAL AND OBTURATOR NEURITIS*

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The associated muscular atrophy in joint conditions due to disease or injury is a fact early and frequently commented upon. Allen[1] mentions references to this in the days of Hippocrates, but no serious efforts to explain the conditions found were made until the early part of the nineteenth century. John Hunter, in 1776, thought that muscular atrophy was due to some sympathetic relationship between the joint and the muscles involved, and Nelaton, in 1837, attempted a somewhat similar explanation. Barker[2] gives America the honor of first directing the attention of the medical world to the condition through the work of John Kerley Mitchell, 1831-1833.

Brown-Séquard in 1860 gave reflex irritation, acting through the vasomotor system, as the cause of the atrophy following joint disease. Sabourin in 1873, on the other hand, thought it was due to the direct extension of inflammation from the diseased joint. Vulpian's idea was that it was due to a reflex influence exerted on trophic nerve centers in the spinal cord. Charcot later supported this opinion. Valtat and Dide, in 1877, experimentally produced arthritis and studied the subsequent muscle changes. Conclusive proof of the correctness of the reflex theory was furnished by Raymond,[3] in 1890, who in a series of experiments on dogs, set up arthritis in both knee joints by injections of nitrate of silver and by cautery, cutting first the posterior roots of the lower lumbar and upper sacral nerves on one side. He found muscular atrophy very marked on the side with the nerve roots intact, while on the side on which the roots had been cut, the muscles either did not atrophy at all, or did so only to a very slight extent. He explained this difference by the inability of noxious influences to travel through the posterior roots into the spinal cord and act on the trophic centers in the anterior horn on the side on which these posterior roots have been cut; on the side with the roots uncut, atrophy necessarily occurred. Duplay and Cazin, in 1891, repeating Valtat's experiments, found the muscle changes consisted only in a simple decrease in the size of the fibers. Hoffa, in 1892, repeated Raymond's experiments and confirmed his results.

Klippel, in 1888, had reported some changes in the spinal cord in arthritic muscular atrophy. None of the observers had been able to discover anything abnormal in either the spinal cord, or the nerves or muscle nerves. Forsbrook[4] in a dissertation on osteo-arthritis, is of the opinion that the muscle atrophy is contingent, first, on the discontinuance of physiologic activity necessitated by the diseased state, and second, on a neuritis following the affection of the articulation. Gower points out that disuse alone cannot explain the atrophy because the results of disuse are generally trifling and tardy, and moreover that other muscles of the limb of which movements are suspended to some extent, show no atrophy. In 1898, Achard and Levy reported finding, in this disease, atrophy of cells of the anterior horn of the spinal cord. Mignot and Mally[5], in 1900, as a result of their experiments on dogs, found on the side of the muscular atrophy, a diminution of the number of cells in the anterior horn throughout the lumbar portion of the cord.

Allen repeated Raymond's experiments

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*Read at the Neurological Conference, Vanderbilt Clinic, March 28, 1921.

and used the finer histologic methods introduced since Raymond's time. He found, as soon as the inflammatory reaction following the injection of the irritant had subsided, in cases without root-cutting, a diminution in size of the muscles on the extensor surface of the limb. In some cases as much as 6 cm. difference was noted in the thighs of the two sides. Faradic irritability was retained but was slightly reduced on the affected side. There was simple reduction in size of the fibers, the space between them being greater than normal. The fibers preserved their striations and neither increase in muscle nuclei, nor any pathological conditions in the nerve endings was found. The peripheral nerves showed no change by either the Weigert or Marchi methods.

The atrophy was most marked on the side with intact nerve roots. As the animals were very much emaciated the muscle fibers were smaller on both sides with equal absence of change in the nerves, with the exception, of course, of the ascending degeneration on the side of the spinal cord on which the posterior roots had been cut. The lower part of the spinal cord, particularly the fourth and fifth lumbar segments, in the former of which Parhon[6] located cells of origin of anterior crural and obturator nerves which supply the anterior muscles and the knee joint, was carefully examined, but not one cell could be found which showed axonal degeneration. The large cells of the anterior horn were carefully counted and differences in the two sides from 1.3 to 7 per cent were found, but a further count of all the anterior horn cells gave figures essentially the same. This, to Allen's mind, raised the question of the possible normal variation in the number of cells of the two sides of the spinal cord.

Nathan[7] in 1916 pointed out the association of symptoms referable to the nerve system with both acute and chronic joint disease. He deprecates the fact that most writers are content to follow Charcot in his view that neural symptoms are due to the reflex influence of the arthropathy. He concedes that the atrophy and hypertonia may properly be attributed to reflex action, but adduces evidence to show that the cases of muscular atrophy and spasticity, vasomotor changes and disturbed sensation, are directly associated with the underlying morbid process. He has found in acute stages of polyarthritis spastic atrophic phenomena, transient in some, more or less permanent in others, evidently not of reflex origin. In some cases he found only neuromuscular phenomena; in others symptoms referable to irritation of sensory nerves—hyperesthesia, radicular and lancinating pain, and occassionally vasomotor and trophic skin disturbances (localized hyperhidrosis, glossy skin and edema) were present. In many cases he found muscular atrophy and spasticity with the joint disease insignificant, the disability due to muscle atrophy and permanent muscular shortening. His experimental work points to the symptoms as being due to radicular or spinal irritation or compression, caused by the altered mechanical condition in the epidural space and intervertebral notches, with no actual involvement of the nerve system. He emphasizes the general infections which give rise to these conditions, often overlooked in the history of the case and therefore giving the case the appearance of a chronic progressive disease. His experiments on dogs, which he inoculated with streptococci, showed vertebral changes, endosteal subperiosteal inflammation, concomitant with sterile intra-, peri- and para-articular exudates. In the spine this exudate which involved the epidural space and vertebral notches was of such a nature that it must perforce have caused spinal root irritation or compression. In some the exudate was finally absorbed, in others, permanent connective tissue changes occurred. The spinal lesions in the dogs experimented upon were the same as in the peripheral joint lesions. He reasons thus: Since the peripheral joint lesions in man and dog are absolutely analogous, and we have no reason to believe that the bones and peri-articular structures of the spine in man react differently to deleterious influences than do those of the skeleton generally, we may assume that the vertebral and epispinal morbid conditions in man are analogous to those demonstrated in the dog. Hence he concludes that the neurological symptoms described as associated with arthritis point to the involvement of the spinal column in the morbid process. The vertebral changes, consisting of endosteal and subperiosteal inflammation, have led to epidural and perispinal exudations causing irritation or slight spinal compression. These changes may or may not be permanent, differing in distribution, intensity and permanency depending on the distribution, intensity and sequelae of the epidural and perispinal processes. The infection need not necessarily be of a suppurative character and may or may not produce nerve symptoms, such a result being due to difference in location, not to difference in the morbid condition.

The following case is interesting both because it bears on the questions discussed above and because of simulation of an anterior crural and obturator neuritis. Were it a typical case of neuritis, we could supply a rational explanation of the muscular atrophy on that basis, but the clinical aspect of the case warrants only the diagnosis of muscular atrophy, secondary to arthritis of the left hip, with paresis or partial paralysis of the muscles supplied by the anterior crural and obturator nerves.

History. A. F., male, of German extraction, aged fifty-six, a butcher by occupation, widower, careworn and apparently in pain, presented himself at the nerve clinic of the Post-Graduate Hospital, Neurological Department, on January 17, 1921, with the following complaints: pain in the left groin, pain in the left hip region, pain in right ankle region and pain and difficulty in walking. His first symptoms dated back three years and began with slight pain in the left thigh anteriorly. The onset was gradual and the disease became continually worse. At that time, his pain occurred about three or four times a week and lasted but a few hours. It was of a sharp shooting character going right through his leg. The course of the pain was from the left groin to a little below the knee-cap of the left leg. He would have to discontinue his work, go home, and lie down in bed. The next day the pain was generally better. Perhaps he would be able to work from morning until evening on some days, but in the evening the pain would become very severe and the only comfort he could secure then would be to lie quietly in bed with his legs stretched out. No form of medication, no amount of hot applications of various sorts, would afford him as much relief as getting to bed. At the onset of his difficulty, he would limp a little, but no considerable difficulty in walking was experienced until about one year ago when he became conscious of the fact that his left leg was weaker than the right, the main weakness being in the upper thigh. This progressively increased until to-day it is difficult for him to get along without a cane. He also noticed a difference in the size of the two thighs about six months ago. He prefers standing most of the time. He finds no comfort in sitting. When sitting, he selects the edge of the chair and sits down sidewards on the right buttock. When sitting and stretching his left foot (i. e., hips partially flexed) the pain begins in the anterior thigh region. When lying down it eases up a little. In getting up he experiences more difficulty. Sometimes after struggling to his feet, he can hardly walk. He often has to hold on to the bed or use a chair to reach the cupboard or sink. The continuous pain is of a burning or tearing nature, occasionally it feels shock-like, especially after a strain, pull or misstep. It is severe in rainy or cloudy weather, and he claims to be able to foretell the character of the weather by the severity of his pains. During the actual storm or rain the pain becomes a little less severe. He further complains that the left leg and foot are colder than the right. About two years ago he had a peculiar sensation as of ants creeping up and down the leg. It lasted for a few months and though more severe on the left, was also present on the right thigh and leg, lasting one to three quarters of an hour at a time.

His past history is practically negative. To his knowledge he was normally born, breast-fed, had the usual diseases of childhood and was particularly subject to sore throats. About fifteen years ago, he had an attack of pneumonia which left him in very poor health. In one year he had recovered his former state of good health and has enjoyed it until three years ago. He denies any venereal infection, and denies lues by name and symptom. He was a very heavy meat eater, eating large quantities three times a day up to the onset of his illness. He never suffered a typical attack of gout nor had any trauma or temperature during the course of his illness. To his recollection there has never been any trauma to his left hip. He was accustomed to several whiskeys and some glasses of beer, up to the onset of his illness. Since then he only imbibes an occasional glass of beer and has four cups of coffee or cocoa per day. His sleep is very poor, he is restless and nervous. His appetite is good, thirst moderate; urinary history negative, voiding normal amounts with an occasional nocturnal urination; bowels regular; potency and habits are normal. His sexual life is limited. He had been married only five years when he was widowed. His wife had no pregnancies during that time. His family history is negative on both his paternal and maternal sides, particularly in reference to diabetes and nervous affections. He has weighed at best 190 lbs. To-day he weighs 150 lbs. He has had no operations, and gives no history of injury, fracture, dislocation or disease of the vertebrae. An examination on March 25th gave the same clinical findings as below described. His history then referred to pain directly at the left hip, running to the left groin for the past two months.

Examination. Abnormal attitudes and deformities: He holds his left leg stiffly, bearing his weight on the right.

Gait. He walks in such a manner as to avoid bending his knee, his left leg held stiffly has a pendulum motion. The patient states that he steps off the car backwards and when going down stairs does similarly.

Coordination. Patient is right-handed and performs the equilibratory tests, finger-to-nose, and finger-to-finger, normally. The heel-to-knee test on the left is painfully performed but shows no ataxia. There are no adiadochokinesis, dyssynergia or dysmetria. Skilled acts are normally performed. No abnormal involuntary movements are noted. Deep reflexes are normal. Attention is called to the fact that his knee-jerks are marked 2. = normally. No ankle clonus was elicited. A possible difference in the cremasteric reflexes is noted, the left being slightly less active. Of the superficial reflexes, the abdominals of the right side are not elicited. This perhaps may be explained by the laxity of the abdomen on that side and also by the presence of an interesting tumor, situated in the upper and lower abdominal quadrants. It is fusiform in shape, about 6 inches in length, 2½ inches in breadth, feels like fat or omentum, moves with respiration and is not attached to the skin. It seems to be attached to the external border of the left rectus muscle, and gives an impulse on coughing, but not more than the impulse recorded on the right side.

Muscle Strength. The patient, it should be noted, has been a powerful man and was accustomed to lifting heavy weights of meat. His muscle strength over trunk and upper extremities seems to be normal. In the lower extremity the left thigh muscles are affected. Flexion of the thigh on the left is performed with difficulty and pain, evaluated at 5. = movement limited in extent. Extension and abduction are apparently normal. Adduction is slightly impaired. Flexion of the leg is normal. Extension of the leg is impaired, graded 5. The feet and toes seem to be normal in plantar extension, and plantar flexion; for abduction and adduction this also holds true.This seems to point very markedly to impairment of the motor power of the iliopsoas, sartorius and quadriceps muscles supplied by the anterior crural nerve, and to a lesser degree to involvement of the pectineus and the abductor muscle group, the longus, brevis, and magnus, supplied by the obturator nerve. There is present, besides the marked atrophy of the anterior thigh muscles, an associated hypotonus and softness of the muscle structure.

Muscle Status. The measurements of the thigh at two points of greatest circumference show—right 18½, left 17½ inches; at another point—right 19½, left 18¼ inches. Electrical test shows diminished excitability and no changes in polarity in the anterior crural and adductor group muscles.

No reaction of degeneration is present in any muscle.

No abnormal associated movements are noted.

Sensory. The general sensory examination is practically negative. A very slight hypesthesia over the internal surface of thigh and leg on the left side, measured by the Strong esthesiometer, shows ¼ inch variation from the normal (which corresponds to the area of the saphenous longus). There is no sensory disturbance over the pubic region and sexual appendages.

Cranial nerve examination discloses no pathological findings. In tests for intelligence, emotion, attention and memory he responds normally.

Systemic Examination discloses that the left leg and thigh are slightly colder than the right; the skin appears smooth and glossy. His *cardiovascular condition* shows his pulse rate 80 per minute; the heart sounds normal. His blood pressure is 150/80. There is present a pulsation of both dorsalis pedis arteries and a marked pulsation of the left femoral artery. The respiratory tract is negative. *Gastrointestinal* tract discloses the hernia or tumor mentioned above, with difference in opinion as to its nature. His teeth are fair, showing some pyorrhea. Genito-urinary examination discloses inguinal hernia. Skeletally he shows the left foot slightly flat. Rectal examination is negative.

The Surgical Department of the Post-Graduate Medical School and Hospital reported on the abdominal tumor as follows: "A positive diagnosis seems to be impossible. The division is between a lipoma of the abdominal wall and a hernia into the abdominal wall."

Examinations of urine 1/25/21 and 2/19/21 were negative. Wassermann test by the Department of Health, N.Y.C., 2/21/21, was negative. Roentgen-ray examination shows the bodies of the vertebrae narrow in their vertical diameter, and proportionately wide. The vertebral discs are proportionately widened. There is no evidence of erosions or necrosis and the above described condition is considered probably congenital. Radiographic examination of the left hip shows the characteristic changes of a hypertrophic osteo-arthritis. The interarticular space is narrow, the articular border roughened and sharpened. Since this condition does not appear to exist on the right side, it is suggested by the *X*-Ray Department that the condition on the left side is due to an old injury.

CONCLUSIONS

Conscious of the fact that some of the factors that warrant the diagnosis of anterior crural and obturator paralysis are lacking, we feel that many points have been brought out in favor of the involvement of the muscles supplied by these nerves. Among these are the weakness of the iliopsoas, sartorius, and quadriceps, atrophy of anterior thigh muscles, the electrical changes, the history and the location of sensory findings. The cause of the primary involvement must remain in doubt. It is certain that there is no history of trauma to the hip, nor any fracture or dislocation of the vertebral column, nor of any rectal condition that is palpable. The question of chronic alcoholism and more particularly of a gouty or rheumatic diathesis, favoring the left thigh through some occupational or other factor, making the left thigh the "locus minoris resistentiae," must be held in mind. On the point of pathology the question arises whether a very slow perineuritis or interstitial neuritis had not taken place, resolving before radical structural changes had occurred in the parenchyma. The symptoms are mainly motor, but there is a possibility that the paresthesia of two years ago was the condition called "meralgia paresthetica." On the point of diagnosis, it would be very difficult to differentiate the muscle condition from a beginning progressive muscular atrophy save for the active knee-jerk. From chronic poliomyelitis it is differentiated by the fact that the knee-jerk is present. The absence of fibrillary twitches and the electrical changes also speak against that condition. Tabes dorsalis and dementia paralytica often give hypertrophic changes in the knee and ankle joints, but can be readily excluded in this case. Syringomyelia gives hypertrophic enlargement of the extremities of bones with muscular atrophy but the location and lack of sensory dissociation speak against this condition.

Some cases of psychoneurotic arthralgia are associated with a higher degree of muscular atrophy than is commonly seen from simple inactivity, but the psychological make-up and definite organic findings readily exclude this possibility. There remains the so-called "reflex atrophy" to explain the muscle affection. Charcot spoke of the influences from the inflamed joint as producing a sort of inertia or stupor of the cellular elements, and others have assumed a so-called "dynamic alteration" in the cells of the anterior horn. Lorenz[8] holds that a muscle maintains its nutrition principally because the peripheral neuron is intact. There are no special trophic nerves as far as is known and the nerve centers do not spontaneously exert a trophic influence, but must be stimulated from the periphery and from the center in order to do so. The cutting off of voluntary influences may thus result in a diminution in size of the muscle, as in the atrophy sometimes observed after a cerebral lesion. Similarly in the muscle atrophy following joint diseases or injury we may analyze several factors. First, the limb is held in a fixed position more or less and produces atrophy from disuse. Second, the greater or less reduction in the number of the habitual stimuli to the nerve centers takes place and the muscle is apt to suffer. We have no history nor findings pointing to effusion into the joint, so we may eliminate the overstretching of the muscle and pressure which may have been a factor. There is a possibility that the toxic substance produced in the inflamed joint may have an unfavorable action on the muscle, directly selecting certain groups of muscles through their offering a point of least resistance. Direct extension from the joint may be suggested, but no trace of such activity can be found. Hilton's law points to the similar innervation of the joint and the muscles moving that joint. The muscle tonus of the quadriceps extensor depends on impulses which are carried back to the spinal cord by the afferent fibers of the anterior and obturator nerves which include fibers from the knee-joint. In our case there is a flabbiness and lack of tone of the extensor muscles. The tone, however, is not entirely absent, for the knee-jerk is present; in fact is fairly normal. This is due to the fact that impulses arising from the synovial membrane of the knee-joint can pass to supply the muscle partially. Though the reflex influence from the inflamed joint acting on the nerve centers in the spinal cord may not completely satisfy us as being a factor in the muscle atrophy, nevertheless, we cannot totally disregard it.

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TWO CASES WITH SIGNS OF INCREASED INTRACRANIAL PRESSURE PRESENTING UNUSUAL ROENTGEN FINDINGS IN THE CRANIUM

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The writer feels warranted in reporting these two cases not only because of their unusual clinical interest and the noteworthy roentgen findings, but also because of the practical consequences incident to the diagnosis.

CASE I. *An instance of cortical epilepsy is described with extensor movements of the left upper and lower extremities and left face, associated with an unusual roentgenogram of the calvarium (intracranial osteoma?)*. The disappearance of all symptoms *without operative intervention,* the results of physical and roentgenographic re-examination three months after the onset, together with the therapeusis employed, are of unusual interest.

The patient is a white man, twenty-five years of age, whose history follows:

Habits. Irrelevant. No alcoholic or drug addiction.

Family History. Negative. No one in the family has had an illness similar to the one afflicting the patient.

Previous History. He had "yellow jaundice" seven years ago, malaria twice while in service in the Philippines; "rheumatism" in the left ankle-joint of unknown cause, eight years ago. *No history of head injury.* Venereal disease denied.

Present History The patient was admitted to the U. S. A. Base Hospital, Camp Merritt, N. J., December 6, 1918. For the previous nine days, the patient had been subject to attacks of tonic muscle-spasm. The onset was sudden, with no ascertainable cause. At first the spasm was limited to the extensor muscles of the left arm, forearm and hand. About ten hours after the onset, the extensor muscles of the left thigh also became involved; the next day, the muscles of the left face were implicated (involuntary closure of the left eye—orbicularis palpebrarum). The facial involvement lasted just one day.

No pain, unconsciousness, dizziness, headache or mental confusion attended any of these attacks, which were of very frequent occurrence (five to eight an hour).

At the onset, during severe attacks, some speech difficulty occurred, which was not in the nature of an aphasia or anarthria; this has since disappeared. The attacks occur at night and frequently awaken him. There are no other subjective complaints. He has never had anything like this before.

Physical Examination. Special senses, negative. Skin and mucous membrane, negative. Two interesting congenital anomalies are present; one is a peculiar disposition of hair on the forehead, i.e., instead of the usual convex line which marks the beginning of the hair on the forehead, there is a peak or promontory of hair jutting forth in the middle of the forehead; making a V-shaped border of the hair line. The other is a malformation of the tragus of the left ear, —upon the usual structure there is superimposed a small supernumerary formation.

Vascular System. Negative. Blood pressure, S. 146; D. 84. During an attack it was S. 156; D. 90.

Heart and Lungs. Normal.

The temperature, pulse and respiratory rates were normal during the sixteen days constituting the first period of observation.

NEUROLOGICAL EXAMINATION

The Attack. The patient has frequent, repeated attacks of tonic muscle spasm. Without any aura, the extensor muscles of the left arm, forearm and fingers become involved in a tonic spasm, followed very quickly by involvement of the extensor muscles of the left thigh. The extensor muscles of the left arm and forearm stiffen, the fingers straighten out, and become hyperextended, and the left thumb becomes adducted; a gradual outward rotation of this extremity through an arc of about 45° also takes place. Following quickly upon the upper extremity, the quadriceps extensor of the left thigh goes into a tonic spasm and the leg becomes involuntarily extended on the thigh. No facial involvement occurs; during the seizure he is able to move all the muscles of his face with ease. The entire seizure is over in six to ten seconds. Following it the patient complains of a distinct numbness in the parts affected, less in the thigh than in the forearm. *He has from six to eight attacks an hour.*

Motor. The strength in the left triceps, deltoid and in left-hand muscles (grip) is very slightly diminished. The patient is of magnificent muscular development, revealing no other motor disturbances.

Sensory. No objective disturbances.

Reflexes. Those of the forearm, biceps, and triceps are quite exaggerated, more so on the left side. Both Achilles tendon jerks are increased and equal; both knee-jerks are exaggerated, the left distinctly more so. No ankle clonus; no Babinski or allied reflexes; no Hoffman digital reflex; no abdominal reflexes. Right cremasteric, present; left, absent.

No Romberg. No astereognosis. No adiadochokinesis.

Cranial Nerves. Eyes: the pupils are regular, equal, round, and react to light. There is slight lateral nystagmic twitching of the eyes. Fundi: right, normal; left, shows a myopic refraction error. Media: normal. No hemianopsia. Hearing: normal. No cranial nerve abnormalities.

Serology and Laboratory Data. Lumbar puncture revealed a clear fluid under very little pressure; the fluid stopped running after about 1 cc. was obtained, this amount being entirely consumed in performing the Wassermann reaction which was negative.

Blood Wassermann. Negative.

Blood Count. Red corpuscles, 4,400,000; white corpuscles, 9,600.

Differential Counts

Polymorphonuclears	
Neutrophiles	70 per cent
Eosinophiles	6 per cent
Small mononuclears	18 per cent
Large mononuclears	5 per cent
Transitional leucocytes	1 per cent

Repeated Urinalysis. Negative

Roentgen-Ray Findings. A mass having almost bone-like density extends downward and forward just anterior to the junction of the frontal and parietal bones. This mass appears to be dipping into the longitudinal fissure, and is about 4½ cm. deep and 1½ mm. in width.

Subsequent Course. Re-examination December 17, 1918, reveals the same findings that the former examination disclosed, with the following exceptions: bilateral ankle clonus; definite nystagmus on looking to the right, none to the left. Recently the severer seizures have been associated with tonic spasms of the left face.

After December 20th, the seizures gradually became less frequent, until by December 23rd they ceased altogether. At this time he was transferred to another military hospital and was not seen again until March 21, 1919. In the three months (December to March) he had had no seizure and had been in excellent health. During that period, his blood and spinal fluid were subjected to the Wassermann test, with negative results.

NEUROLOGICAL RE-EXAMINATION
(March 21, 1919)

Motor. No weakness, paralysis, or atrophy anywhere.

Sensory. No objective or subjective disturbances.

Reflexes. There is a distinct increase of deep reflexes on the left side, with absence of superficial reflexes on the same side.

Knee-Jerks. Both active, left more than right. Achilles tendon jerks: both active, left more than right. No Babinski, Oppenheim or Chaddock. Triceps, biceps, and forearm reflexes active; greater on the left than on the right side. Abdominal reflexes: right, diminished; left, absent. Cremasteric: right, active; left, absent. Corneal reflexes: normal.

Cranial Nerves. Eyes: pupils equal, react actively to light. Fundi: essentially negative. No hemianopsia. No nystagmus.

A peculiar and marked thinning of the hair has occurred in the left temporal region.

The roentgen-ray findings are the same as previously noted.

THERAPEUSIS

Before admission to the hospital he was given 15,000 units of tetanus antitoxin. From December 7th to December 13th, he was given sodium bromide, .65 gm., every four hours, and occasional doses of morphine sulphate, .008 gm. On December 15th, potassium iodide was begun in 1 gm. doses, three times a day, increasing .06 gm. a day until December 22nd, the date of his transfer to another hospital. On December 16th and 21st, he was given .04 gm. of mercury salicilate in albolyn intramuscularly.

Reference has been made to the fact that on December 20th his symptoms began to abate, and in three days had entirely disappeared.

Was this result the effect of specific treatment on a luetic condition, or with the negative serology and history in mind, were the improvement and the therapeusis merely coincidental circumstances? Are we dealing with an intracranial osteoma with a luetic process (namely syphilitic meningitis) engrafted upon it, with the osteoma latent and symptomless, until the luetic process became active? Or again, in view of the visible congenital abnormalities mentioned, have we a congenital condition (namely, teratoma) with or without a luetic pathology added?

In view of the successful issue of the case, such questions are very difficult to answer, and an exact diagnosis is impossible at present.

Inasmuch as this patient was scheduled for an operation for intracranial neoplasm, the unforeseen but fortunate turn taken by the case seemed all the more remarkable at the time.

Schüller in his excellent monograph[1] and Sippel[2] have both discussed the subject. The former, speaking from an unusually large experience, states that "apart from tumors of the bones of the face, roentgenographic pictures of tumors of the bones of the skull are quite rare." He cites 4 cases: 2 frontal, 1 orbital, and 1 bregmatic osteoma (similar to the one herewith described) ***in none of which convulsive phenomena occurred.*** He further states that "circumscribed osteomata, . . . according to their site, have a course free from symptoms or are associated with severe cerebral, nervous or ocular disturbances."

CASE II. ***Paralysis of the first six cranial nerves on the right side complicating fracture of the right side of the sphenoid bone, and adjacent floor of the orbit. The very slow involvement of these nerves, the negative serology, and the x-ray findings constitute an interesting clinical picture.***

The patient is a male, white, forty-eight years of age.

Habits: Drinks very little alcohol, never intoxicated. No drug addictions.

Family History. Negative.

Previous History. Scarlet fever at fifteen years of age. Occasional attacks of vertigo and syncope. Venereal disease denied.

Present History. In January, 1916, while playing baseball, he was struck by the ball in the region of the right eye. At the time he had a severe traumatic epistaxis, and was unconscious for a short time; he states he was "pretty well knocked out." Six months later (June, 1916) severe pains on the right side of the nose and adjacent right malar region appeared. These "neuralgic" attacks became rapidly worse, involving also the right forehead.

Two operations were then performed upon the right side of his nose, directly after which the sharp pains disappeared for six weeks, then returned again, localizing "in the right eye." At this time he noticed that the visual acuity of his right eye was becoming impaired. While this diminution in vision was progressing, he noticed a loss of sensation in the skin over the right malar region.

In May, 1918, while going through some "gas tests" at Camp Greene, he lost the vision of his right eye.

While the visual defect was progressing in the last two years, the "neuralgic" pains were increasing in severity. After an attack of pain, he states, the right eye would "clear," but he noticed that actual vision was getting weaker and weaker in that eye until it disappeared entirely in May, 1918. In the following two months the pains described diminished, until June, 1918, when the pain disappeared; in place of it there was numbness.

After the injury (January, 1916), he was subject to attacks of nausea for about six months; at no time did he vomit.

NEUROLOGICAL EXAMINATION

Motor. Marked atrophy and loss of power of right masseter, pterygoids, and temporal muscles (motor portion of mandibular division of trigeminal nerve).

Sensory. Complete loss of all forms of sensibility in the cutaneous and mucous membrane distribution of the right trigeminal nerve. Loss of taste (sweet, sour and bitter) on the anterior two-thirds of the right half of the tongue (involvement of chorda tympani branch of trigeminal). Anesthesia of right cornea. Loss of smell on right side.

Reflexes. Knee-jerks, Achilles tendon-jerks, abdominals and cremasteric reflexes are equal and active on both sides. The corneal reflex is absent on the right.

Cranial Nerves. Eyes: pupils, unequal; right, semi-dilated and larger than left. The right pupil is fixed and does not react to light, accommodation or convergence; the left reacts well. Fundi: right, optic nerve atrophy (post-neuritic type) with complete blindness; left, normal; vision 20/20. The right eye is somewhat sunk in (enophthalmos), and there is some ptosis of the right upper eyelid (cavernous sympathetic involvement). The right superior rectus, external rectus, and superior oblique muscles are paralyzed (incomplete involvement of the oculomotor and complete involvement of the trochlear and nerve to the external rectus). The seventh to twelfth cranial nerves are not involved.

Heart, Lungs and *Abdomen.* Negative; temperature, pulse and respiratory rates, normal for the period of observation.

Laboratory Data. Urine: negative. Spinal fluid: pressure normal, appearance clear, no clots. Globulin: negative. Wassermann: negative. Cell count: three.

DISCUSSION OF FINDINGS

Roentgen-Ray Findings. Very much enlarged posterior clinoid processes of sella turcica, with some encroachment on pituitary fossa. An irregular line, suggesting an old fracture into the right greater wing of the sphenoid bone, continuing into the floor of the right orbit is present. It also extends along the nasal side of the orbit into the right frontal sinus, which is somewhat clouded, giving the appearance of a peculiar thickening in the right frontal sinus.*

It will be recalled that the third, fourth and sixth, together with the ophthalmic and superior maxillary branches of the fifth cranial nerves, traverse the cavernous sinus, in company with important sympathetic fibers; that the optic nerve leaves the interior of the skull through the optic foramen; that the posterior part of the olfactory tract is situated near the optic nerve; and that the inferior maxillary (mandibular) branch of the fifth leaves by means of the foramen ovale.

In the region of the cavernous sinus and the adjacent region of the posterior pole of the orbit, they are all grouped in fairly close

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*Unfortunately this interesting *x*-ray plate was broken after the patient was lost track of, and consequently no print could be made of it.

contact. Here then is the site of their involvement in a pathological process.

DISCUSSION OF FINDINGS

Attention is called to the involvement of the cavernous sinus sympathetic fibers.

The cervical sympathetic fibers innervate the involuntary musculature, engaged in elevating the eyelid, projecting the eyeball forward (so-called Mueller's muscle) and dilating the pupil; these sympathetic fibers (preganglionic) enter the skull through the foramen and canal with the internal carotid artery, traverse the cavernous sinus and enter the orbit through the sphenoidal fissure; these fibers then form the sympathetic root of the ciliary ganglion. Post-ganglionic fibers now course as the short ciliary nerves to the above-named muscles.

The involvement of this sympathetic tract, whether in the neck, as in the case of stab wounds, or in the cavernous sinus, as in this case, is evidenced by the paralysis of these three involuntary muscles, which, thrown out of function, allow the eye to drop backward into the orbit, the upper lid to sag downward, and the pupil to become small or miotic.

In this case, however, the fixed right pupil has both its dilator muscle fiber (depending upon the above sympathetic innervation) and the constrictor muscle paralyzed. The latter muscle is innervated by the cranial sympathetic fibers accompanying the oculomotor nerve.

It is obvious that the sharp neuralgic attacks in the right malar and forehead regions were dependent upon irritation of the intracranial portions of the maxillary and ophthalmic divisions of the trigeminal nerve (a type of tic doloureux), which preceded the destruction of these fibers.

As to the nature of the pathology, the history of brief unconsciousness, severe nasal hemorrhages following the accident, the very slow involvement of the first six cranial nerves, and the *x*-ray findings, make it seem most probable that we are dealing with a process consequent upon fracture of the right side of the sphenoid bone and adjacent floor of the orbit. Organization of blood-clot with formation of fibrous tissue might readily account for the slow involvement of the cranial nerves. On the other hand, beginning osteoma formation in this region is to be considered.

Of peculiar interest in this case was the fact that the condition had been regarded as a basilar meningitis of syphilitic origin with the unilateral implication of the cranial nerves as a part of the picture. As this soldier was about to be discharged from the army after some twenty-five years' service, such a diagnosis would have robbed him of a disability pension to which he was justly entitled.

The rarity of this clinical syndrome appears in the fact that the writer was unable to find a similar case history recorded.

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ANGIOSARCOMA OF THE LEFT SUPERIOR PARIETAL LOBULE WITH CORTICAL ANESTHESIA

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The saying attributed to Hippocrates, "Life is short and Art is long—the Occasion fleeting, Experience fallacious and Judgment difficult," probably bears with greater cogency and appropriateness upon the differentiation of intracranial neoplasms than upon any other field of medical diagnosis. Failure so often is the only reward in cerebral localization that it is, perhaps, well for us to place on record instances in which success has been the result. This case report presents a number of points of interest and the clear-cut nature of the physical findings and the interesting correlation of the pathological condition with the symptomatology, make a report of the case worth while.

The patient was first seen at the Vanderbilt Clinic by Drs. Strong and Sands in December and was to have been presented at one of the earlier departmental conferences, but the delay occasioned by the holidays increased the anxiety of his family and resulted in his appearance at our office. He was referred to the Neurological Institute where, after a careful preparation, he was shown at one of the morning conferences, at which time definite diagnoses were made and the probable location of the neoplasm indicated.

The patient was admitted to the Neurological Department of the Vanderbilt Clinic on December 8, 1921, and was thoroughly examined by Drs. Strong and Sands. The history and physical findings are as follows:

A. N., aged sixteen. Born in America. No occupation at present. Vanderbilt Clinic Case No. 33,557.

The complaint on admission was "pounding pain at the right side of the stomach, traveling up to the brain and down the right arm to the finger tips. Sometimes I faint; sometimes I am rendered immobile, and cannot see then."

In other details, the history is similar to that which we obtained, and which will be later given in detail.

PHYSICAL EXAMINATION

Abnormal Attitudes and Deformities. None.

Gait. Normal.

Coordination. Equilibratory: Standing with the feet together, eyes open and closed, normal. Standing on one foot is not so well performed with the right, as with the left foot.

Non-equilibratory: The finger to nose test is normal on the left, somewhat ataxic on the right. The finger to finger test on the right is somewhat ataxic especially with the eyes closed, the same result being present in the finger to thumb, the ataxia affecting particularly the little finger. There is no pass pointing. Adiadochokinesis is present on the right side. Slight dysmetria is present on the right in pronation and supination movements. With the arms extended in front and the eyes closed, the right arm tends to drift downward and outward. The right little finger tends to abduct on sustained position of the hand, the patient being unconscious of the deviation.

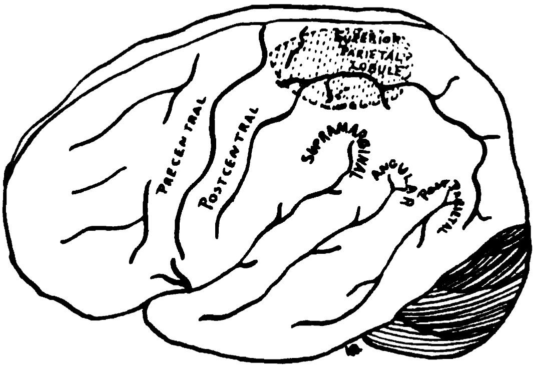

FIG. 1. Lateral aspect of the left cerebral hemisphere indicating the position of the neoplasm.

Skilled Acts. Handwriting is quite good.

Reflexes—Deep	*Left*	*Right*
Biceps	2	2+
Triceps	2	2+
Radial	2	2+
Ulnar	1	1+
Wrist	1	1+
Suprapatellar	2	3
Patellar	2	3
Achilles	2	2+
Ankle Clonus	0	0
Superficial		
Abdominals	Present and active	
Plantar flexion	Active	

No abnormal reflexes were elicited.

Muscle strength is everywhere normal on the left side, the movements at the shoulder, elbow and wrist showing a value of nine which is normal. On the right side the movements of the fingers have a value of seven. In the trunk and legs the strength is normal and equal on the two sides.

Muscle Status. Normal.

Abnormal Associated Movements. No definite abnormal associated movements are present. There is slight tendency to identical associated movement in clenching the fist, left to right.

Sensory Examination. Discrimination is diminished over the right arm, forearm, hand and fingers. Muscle tendon sense is disturbed in the right hand and fingers. Stereognosis is absent in the right little finger.

Cranial Nerves. All are normal in all respects.

Systemic Examination. Some enlargement of the thyroid and tonsils is noted.

Pulse 74; normal.

Blood-pressure: 128/75.

Diagnosis was deferred until further examination, but there was a conviction, on the part of both Drs. Strong and Sands, that they were dealing with a neoplasm involving the superior parietal lobule on the left side.

For the reasons which I have already given, he failed to reappear for a second examination, and instead came to our office on January 19, 1922. His history as obtained at that time is as follows:

His *chief complaint* was convulsions with loss of consciousness.

His *first symptom* appeared in May, 1921, and consisted in what we might call at this time a psychic equivalent, which will be described later.

The *onset* was sudden. The *progress* of the disease has been continuous, the attacks, as they are called, increasing in frequency and severity.

Present Illness. He had been perfectly well up to May, 1921, at which time, while riding his bicycle, he suddenly felt himself lifted off the seat. He dismounted and after standing for a few moments, felt a throbbing pain beginning in the right side of the chest and extending upward into his head. The pain lasted for about ten minutes and did not radiate into the arm or leg. He was fully conscious throughout the entire attack, and after the pain wore off he walked home, the pain disappearing entirely.

In June, 1921, while swimming in the tank at school during lunch hour, he was suddenly called upon to rescue one of his companions, who was in imminent danger of drowning. While doing so, he thinks that the other boy kicked him in the stomach, and a few minutes later while standing by the railing of the pool, he experienced a second attack similar to the first, but of greater severity. This lasted about ten minutes and consisted chiefly in the same kind of pain as outlined above. He had in addition to this, a subjective sensation as though his hand were jumping, but on looking at it, he found that no actual movement was taking place in the hand. His right arm appeared semiflexed at the elbow, the fingers were stiff, and the entire arm felt as if it were held tensely by his muscles. In this attack he lost consciousness for a few minutes, the length of time he does not know exactly, but in any case it was for so brief a period, that he was able to return to his classes at 1 o'clock feeling no immediate bad effects; no headache, nausea or vomiting. There was no loss of sphincteric control.

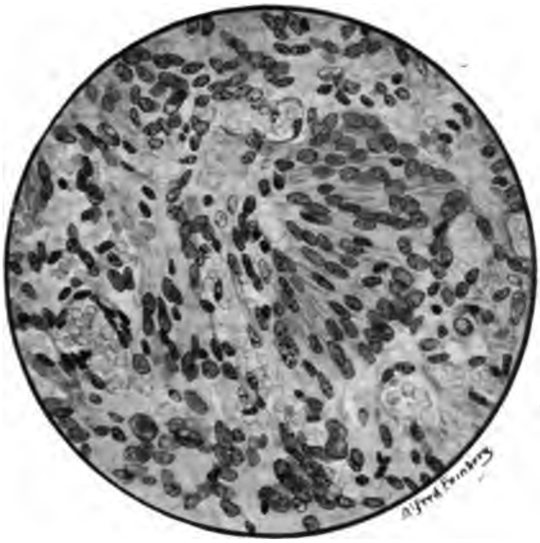

FIG. 3. The microscopical appearance of the tumor. The rich vascularity of the neoplasm is very evident. The character of the tumor is indicated by the bundles and groups of spindle cells which often seem to radiate from blood vessels. In other areas large vesicular nuclei appear embedded in a syncytium. There are a number of these cells the nuclei of which contain vacuoles.

In the interval between these two attacks, there were minor seizures of a similar nature, beginning with the same pain and consisting in a pounding sensation, which at times extended into the right arm. These were very short in duration and during them he never lost consciousness. Often they were so slight that he could continue whatever he happened to be doing, although at first he was very apprehensive when they occurred and would stop and wait until they passed off.

About the middle of June, during the night, the patient had four severe convulsions, all of which occurred during his sleep, although some of them caused a brief period of wakefulness before the patient lapsed into unconsciousness. In one of these his brother saw him kicking around in bed, attempted to arouse him, but failed. He remembers nothing of these attacks and states that there was a great tendency to fall asleep immediately after the attack was over. In none of them was there any incontinence, nor did he ever bite his tongue. A general description of the attacks will suffice to describe each one. They all began with a pain on the right side in the lower part of the thorax, which spread up to the head and outward to the shoulder, then down into the neck, arm and fingers of the right hand. They were characterized by a throbbing pain with a rhythm which corresponded with the beating of his heart. This initial pain lasted for about five minutes and was usually a warning of the approach of a generalized attack. He occasionally had the pain as a warning and then the major attack failed to develop. When the convulsive seizure did follow, he noticed a forcible flexion of the fingers of his right hand, so strong that at times the nails were driven into the palm of the hand and he was unable to extend his fingers. With the contraction of his fingers, there was also a tendency to flex the forearm at the elbow, the arm was adducted and the muscles held in a rigid spasm. On several occasions after the painful aura, the right leg has gone into spasm instead of the right arm. With the severe attacks, following the convulsive movements of the right arm, there was a spread to the entire body musculature and a general convulsion followed with loss of consciousness, cyanosis and moaning.

These attacks have been more or less regular in their appearance, occurring about every three weeks. More frequently than the preceding, but with less regularity, there have been minor attacks, in which he does not lose consciousness and in which the right hand, forearm and arm, or the right leg may go into spasm without any further spread in the manifestations of the attack.

During the first week in July, the patient had two attacks while asleep of which he knew nothing, but of which he was told by his parents. In August, 1921, again in the night, he had one severe attack. After this he felt no ill effects and was not inconvenienced in any way. During the period intervening between August and the middle of September, the patient had only minor attacks which have been already described. On further questioning, he describes the sensation in his fingers "as if someone were holding the fifth finger tightly against the others."

About the middle of September, after considerable exertion in carrying a heavy object, he had a severe attack beginning exactly as before, radiating up into the head and into the right arm, in which the fingers of the right hand were drawn tightly together into the palm, the forearm was flexed, drawn up at the shoulder and the whole arm adducted. Recognizing that an attack was imminent, he called to one of his companions who, seeing his condition, instead of coming to his aid, ran away and left him. In an attempt to let himself slowly to the floor, he did not succeed and fell. In this attack, there were no jerking movements, the arm was held rigid and stiff and he was entirely conscious of everything that was going on about him.

During September, the patient noticed an increasing awkwardness in the use of his right hand, which progressed to such an extent as to render him incapable of playing the piano.

During December, he had one attack followed by headache, which has persisted off and on since then and has been frontal in location. This is the first occurrence of headache noted in the history.

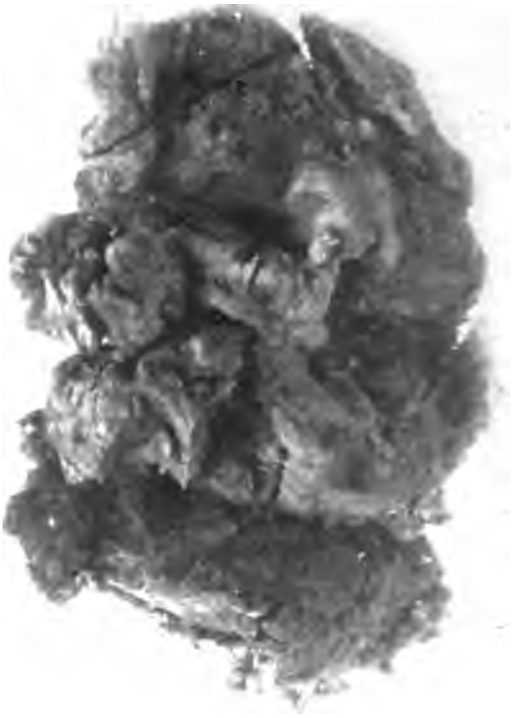

FIG. 2. The shaggy appearance of the under surface of the tumor which showed rather intimate relationship to the underlying brain tissue.

On January 17, 1922, he went home about 3 o'clock in the afternoon, took something to eat and began to read. Suddenly he felt his head turning to the left as though someone were pushing it in that direction. He immediately lost consciousness and fell to the floor in a convulsion. During this attack he vomited. About an hour and a half after recovery, he had a recurrence in which his head again turned to the left, followed by a general convulsion and loss of consciousness. Since this attack, he has awkwardness and loss of the usual dexterity in the use of his right hand, and there is slight dragging in the right foot while walking. This attack was preceded by some dizziness but not by the usual pain aura described before.

Past History. This has been negative in all respects except for the following: He had mumps in childhood, an operation for right-sided mastoiditis in 1915. He had pneumonia in 1915 from which he made a perfect recovery. There has been no history of any diplopia, failing vision, difficulty in hearing or any other abnormality except as noted.

Family History. His mother has had eight pregnancies, three of which resulted in accidental miscarriages. The father and mother are living and well. He has one brother who is in good health and two sisters who are well. There is no similar disease in any other member of the family.

Personal History. The patient is right-handed. He drinks tea and coffee moderately, but does not use alcohol or tobacco. He sleeps well and has a good appetite.

PHYSICAL EXAMINATION

Abnormal Attitudes and Deformities. None noted or described.

Gait. He is slightly unsteady, which is emphasized by efforts to walk on a straight line. The unsteadiness apparently results from an inability to use his right leg accurately. He is able to walk on his toes, on his heels, backwards and sidewise, without any material increase in the unsteadiness.

Coordination. Equilibratory: There is very slight swaying with his eyes open and some increase in this upon closing the eyes. He stands well upon his left foot, but very poorly on his right.

Non-Equilibratory. On the right, the movement in the finger to nose test is unsteady, wavering, and presents oscillations which are increased upon removing the finger from the nose. The test is normal on the left side. The finger to finger test brings out the same wavering movements upon attempts to approximate the fingers. The finger to thumb test is done awkwardly, and there is a tendency for him to keep his eyes constantly on the right hand, in order to perform the movements of the fingers. In the toe to object above patient test, the same phenomena are observed although less pronounced, and in the heel to knee and along shin tests there is the same condition of affairs. There is marked inability to produce rapid succession movements on the right side, and a marked loss in the check element on forced movements of the right arm.

Skilled Acts. There is dyspraxia present in the right hand and his handwriting shows marked awkwardness, irregularities, jerkings and improper measuring in the direction and position of the letters.

Abnormal Involuntary Movements. In addition to those contained in the history, there is a rather coarse tremor of the whole right upper extremity.

Reflexes—Deep	*Left*	*Right*
Pectoral	2	3
Biceps	2	3
Triceps	2	3+
Ulnar	2	2
(with spread to deltoid and biceps on the right side)		
Wrist	2	2+
Suprapatellar	2	2+
Patellar	2	3+
Hamstrings	2	3
Patellar adductor	0	0
Tibio-adductor direct	0	0
Tibio-adductor crossed	0	0
Achilles	3	4

Reflexes—Superficial	*Left*	*Right*
Supraumbilical	Present	Present
Suprapubic	+	0
Upper lateral abdominal	+	+
Lower lateral abdominal	+	0
Cremasteric	+	+
Plantar flexion	Normal	0
Babinski and modifications	0	0

Muscle Strength. Power on the left side is normal. The right arm, forearm, hand, thigh, calf and foot show some diminution in power. The upper extremity is more extensively involved than the lower.

Muscle Status. There is no atrophy, hypertrophy nor any changes in muscle tone.

Abnormal Associated Movements. Not observed.

General Sensory. Touch: Acuity is very slightly diminished over the entire right side, except the face. This diminution can only be elicited by directly comparing the right and left sides. The localization of two points simultaneously applied to the skin, shows that the circles of Weber, to be recognized, require a diameter on the right side twice that on the left. This affects the hand, forearm and arm. Touch localization is considerably disturbed, error in localization on the right being about twice that on the left. Pain: Normal on the face; elsewhere on the right side of the body, a pin does not feel quite so sharp as on the left. The sensation of pinprick is retarded and somewhat more diffuse. Temperature changes are similar to those elicited under pain:—diffusion of the area stimulated, retardation of the appreciation of the test-tube and a slightly diminshed acuity in recognizing changes in temperature. Vibratory sensibility is normal. Muscle tendon sensibility is very much diminished in the right thumb, slow circumduction not being appreciated. The loss in muscle tendon sensibility becomes progressively greater as the little finger is approached, extreme tension being necessary to cause an appreciation of movement in the little finger. Deep muscle sense is diminished in the wrist, somewhat diminished at the elbow and shoulder. In the lower extremity, there is a disturbance in appreciation of movement in the toes, especially marked in the great toe which requires an excursion of 3 to 4 cm. before movement is detected. Stereognosis: Ability to recognize a knife, fountain pen, watch, coin, is lost in the right hand, whereas they are instantly recognized in the left hand. There is acragnosia in the right arm, forearm and hand.

CRANIAL NERVES

Optic and Oculomotor Apparatus. Vision in both eyes is normal and the fields are normal. Fundi; There is a blurring of both discs on the upper and nasal side with a papilledema of one diopter in the right eye. Pupils are equal, 3½ mm. in diameter, round, regular, central in position; no hippus present. The reaction to light, direct and consensual, is normal. There is no Argyll-Robertson pupil. Position of the eyes is normal. There is no strabismus and no limitation of voluntary movement of the eyes. Nystagmus: there are slight oscillatory movements on extreme left lateral gaze. The palpebral fissures are normal.

Fifth Nerve. Normal.

Facial Nerve. There is a slight awkwardness in movements of the right face, especially marked about the angle of the mouth.

Eighth, Ninth, Tenth, Eleventh, and Twelfth Nerves. All normal.

Systemic Examination. Normal.

He was advised to enter the New York Neurological Institute where the following examinations were made.

LABORATORY FINDINGS

Blood Examination

Hemoglobin	86%
Color index	0.9
Red blood cells	4,780,000
White blood cells	5,000
Polymorphonuclear leucocytes	64
Lymphocytes	34
Eosinophiles	2
Non protein nitrogen	28.2
Urea nitrogen	13.2
Blood sugar	0.115
CO_2	48.1%
Wassermann	Negative

Spinal Fluid

Appearance	perfectly clear
Pressure	normal
Cells	none
Globulin	negative
Fehling	positive
Wassermann	negative

Urine

Color	straw
Reaction	alkaline
Turbidity	slightly turbid
Specific gravity	1.016
Albumin	none
Sugar	none
Indican	slight excess
Microscopic examination	Triple phosphates
Phthalein excretion	25% first hour
	10% second hour
	35% Total

Mosenthal nephritic test diet was negative, the urea and chloride excretion being normal.

Roentgen-Ray Examination of the skull showed the calvarium to be even in outline and comparatively even in thickness; no definite evidence of increased intracranial pressure. Intracranial circulation was normal, except for one diploic channel in the mid-occipito-parietal region of the left side, which was slightly widened. Sutures normally united. Frontal sinuses were absent. Ethmoid cells not remarkable. Sphenoid cells were rather irregular in shape, not subdivided to such an extent as is usually seen. Mastoids were extensive on the left side. The sella was small and well rounded. The dorsum extended considerably above the level of the anterior clinoids and inclined slightly forward giving a partially closed-in appearance to the sella. Summary: No definite evidence of intracranial pathology.

SUMMARY

No definite evidence of intracranial pathology.

The course following admission, showed no particular abnormalities. There was some headache, dizziness, and occasional warning of attacks which did not materialize. On one occasion he was nauseated and vomited after a severe headache.

The diagnosis was a neoplasm, probably an endothelioma, situated on the surface of the brain in the left superior parietal lobule. Operative interference was advised and he was transferred to the Surgical Service of Dr. Charles A. Elsberg.

On January 31, 1922, examination showed approximately the same signs and symptoms as enumerated above, except that there was increase in the hyperreflexia on the right side, with the production of a pathological inexhaustible ankle clonus.

He was operated upon January 31, the operation consisting in a left osteoplastic flap. The bony tissue was found to be fairly thick and quite vascular. The dura was very vascular, not discolored; the tension was markedly increased. There were no focal irregularities or changes in contour observed. Puncture of the ventricle was attempted but was not successful.

Situated in the superior parietal lobule, there was discovered a tumor about 5x6x3 cm., which was adherent to the dura over a small area about 3 cm. from the midline. The tumor began at the Rolandic fissure and extended backward into the superior parietal lobule over an area about 6 cm. in diameter. The cerebral convolutions about the tumor appeared normal in color and consistency. The tumor was well encapsulated about most of its periphery, but was not clearly differentiated from the cortex in the deeper areas and there was some destruction of the brain tissue as the tumor was removed. The tumor was not particularly vascular. On account of its relationship to the large veins in the Rolandic fissure, there was considerable hemorrhage after its removal and the patient went into severe shock, for which appropriate treatment was given at the time with a rapid improvement in his general condition. The tumor was entirely removed in pieces and the operative defect was closed in the usual manner. He rallied well from the operation and has shown a continuous progressive improvement ever since.

PATHOLOGICAL REPORT OF THE TUMOR

BY DR. LOUIS CASAMAJOR

Diagnosis. Angiosarcoma. The tumor consists of many small blood vessels with cells arranged rather irregularly about them. Some of these cells are small thin spindles, while others are irregular, rounded, containing large rounded or oval rather faintly staining nuclei. In some of these nuclei, vacuoles appear. These latter cells tend to gather together in irregular rounded masses. Both types of cells are irregularly arranged around blood vessels; there is much free blood in the tissues.

Examination on February 4 showed that there was some improvement in the deep muscle sense in the right hand, improvement being more marked in the pre-axial than in the post-axial portions of the hand. He had regained his ability to recognize a knife, safety pin, and realized that a spoon had a handle and a hollow bowl, but was not able to synthesize these fragments of information into ability to name the object. The reflexes were as before, the ankle clonus was still inexhaustible. The general condition was very good.

On February 8 the papilledema had practically disappeared.

On February 8 the papilledema had continued improvement in his ability to recognize movement and objects. There was still some ataxia present, which was especially marked in the large joints of the right arm and leg.

Following the operation there was no material motor weakness in the right upper or lower extremity, except that immediately following the operation there was a series of rapidly repeated convulsive seizures, four in a period of half an hour, in which the right side was affected with marked convulsive movements of the right hand and there was a paralysis for an hour and a half in the right hand.

Examination made March 8, 1922, showed the following:

Interval History. The patient states that he has improved in every way. He has had no convulsions, no headache, and the only abnormal sensation of which he complains is a feeling of tension between the thumb and index finger of the right hand. He has resumed his piano playing and succeeds very well with the right hand, though not so well as with the left hand.

PHYSICAL EXAMINATION

Abnormal Attitudes and Deformities. There is a scar of a recent osteoplastic flap operation in the left parietal region. At the lower anterior angle of the left parietal bone, there can be palpated a small bony defect. There is no bulging and the bone is solidly in place.

Gait. There is some slight unsteadiness in turning both to the right and left; otherwise the gait is normal.

Coordination. Equilibratory. Normal both with the eyes open and closed. Standing on the right foot is not quite so well done as standing on the left foot.

Non-equilibratory: In the finger to nose test there is some oscillation toward the end of the act, but not on maintaining the finger on the tip of the nose. The same is present in the finger to finger test. The heel to knee test is well done on both sides, but the heel along shin test on the right shows some wavering. The toe to object above patient test is normal on both sides.

Dysmetria: There is some loss of the check element and overreaching in movements performed with the right hand.

Adiadochokinesis is present in rapid succession movements of the right hand.

Skilled Acts. There is an awkwardness on attempting to snap the fingers, although this can be done fairly well with the right hand, and also upon approximating the right thumb to the fingers of the right hand. The speech is normal. The handwriting is slightly better than formerly.

Reflexes—Deep	*Left*	*Right*
Jaw	2	2
Pectoral	2+	3
Biceps	2+	3+
Triceps	2+	3+
Radial	2	3+
Ulnar	2	3+
(On the right side there is an active spread from the ulnar and radial reflexes to the biceps and triceps)		
Suprapatellar	0	1+
Patellar	2+	3+
Achilles	3+	5
Periosteal uncrossed	0	+
Periosteal crossed	oL-R	oR-L

Reflexes—Superficial	*Left*	*Right*
Ciliospinal	+	+
Abdominal	+	0
Cremasteric	+	+
Plantar flexion	+	0
Babinski	0	+
Fanning	0	+
Chaddock	0	+
Oppenheim	0	0
Gordon	0	0
Schäfer	0	0

Muscle Strength. On the right side there is a slight symmetrical weakness in all movements, more marked in the upper than in the lower extremity. Efforts at forced movements in the right hand cause a clonus at the wrist.

Muscle Status. There is no atrophy or any change in contour; tone is apparently normal.

Abnormal Associated Movements. The pronation sign of Strümpell is positive in the right upper extremity; negative in the left arm.

The radialis sign of Strümpell shows a definite increase in the flip of the right wrist dorsally on closing the fingers.

The thumb sign of Klippel-Weil shows more movement toward flexion of the distal phalanx of the thumb, when the clenched fist is forcibly opened on the right side than on the left side.

There is no Hoffman sign present.

The sign of Neri produces a slight flexion in the right leg, upon flexing the extended leg on the abdomen.

There is some elevation of both legs in the trunk-thigh sign of Babinski, but the movement is a little more marked in the right leg. Modification of the trunk-thigh sign of Babinski, with the legs hanging over the table, is positive on the right, negative on the left side.

The coughing sign of Huntington is present on the right, not on the left side.

The complementary opposition sign of Grasset and Gaussel and of Hoover is present; greater downward pressure being exerted by the left leg by elevating the right than vice versa.

The tibialis sign of Strümpell cannot be elicited.

The sign of Marie-Foix is not present.

Sensory Examination. Touch is normal everywhere except for a little hypesthesia on the dorsum of the right thumb and the right index finger. Localization: There is about the same error in the forearms on both sides, but the error in the right arm and shoulder is about twice that in the left arm and shoulder.

Muscle tendon sense is very much disturbed in the right hand, fingers, wrist, elbow and shoulder, but particularly so in the fingers. The same is true of the right foot, but to a much less degree and limited to the toe joints.

Stereognosis: He can now recognize a knife, safety-pin, coin (but he misses the denomination of the coin) and a fountain pen. On the right hand, he cannot distinguish between a square and a round object rolled over the fingers.

CRANIAL NERVES

First Nerve. Normal.

Optic and Oculomotor Apparatus. Vision is normal. Ophthalmoscopic examination shows evidence of a recent papilledema, the right disc is indistinct and blurred except below and laterally; elsewhere there are some radial markings running over the edge of the disc: the veins are somewhat tortuous. In the left eye, the outline is slightly less distinct than in the right eye. There are no hemorrhages. Pupils: The right is 2½ mm., round, regular and central. The left is 3 mm. There is marked hippus present. The pupillary reaction to light, direct and consensual, and on convergence, is normal in both eyes. The palpebral fissures are normal. There is no strabismus. The conjugated eye movements are normal. Nystagmus is not present.

Fifth, Seventh, Eighth, Ninth, Eleventh and Twelfth Nerves. Normal.

Systemic Examination. Normal.

In considering the case as presented, the facts lead one to the diagnosis of an organic lesion in the brain, affecting the superior parietal lobule and manifesting its presence primarily, so far as the examination goes, by disturbances in the cortical association, correlation and synthesis of the primary qualities of sensation as received by the post-Rolandic cortex and transmitted to the superior parietal lobule for elaboration. In regard to the history, the condition is mainly characterized by evidences of cortical irritation manifesting themselves in two types of seizures, one abortive and resulting only in a pain aura and spasmodic contractions of the right arm and forearm; the other, major attacks which develop out of the minor attacks and proceed to a general convulsive seizure, loss of consciousness and the usual post-convulsive habitus. The history and physical examination present the evidence of an expanding neoplasm situated on the left side of the brain, affecting primarily the secondary sensory association centers and secondarily the motor cortex. The disturbances present an adiadochokinesis, a dysmetria and an ataxia which are sensory in character and largely dependent upon faulty appreciation of the position of the muscles in space and the consequent disturbance in the motor pattern as manifested in attempts to produce rapid succession movements and voluntary acts. The reflexes are indicative of pressure upon the pyramidal system passing to the right side of the body, with a general increase in the reaction of the right side reflexes, loss of the superficial reflexes and the presence of some of the abnormal tendon and cutaneous reflexes on the right side. The failure of the great toe on the right side to go into plantar flexion or dorsiflexion upon stimulation of the sole of the foot is indicative of the fact that in the development of the Babinski there is a period, so to speak, of equilibrium when neither dorsal nor plantar flexion results from stimulation of the sole of the foot. The changes in muscle strength indicate a partial interference with the function of the pyramidal tract going to the right side. The sensory disturbances indicate the results of pressure on the post-Rolandic convolution producing a very slight diminution in the cortical appreciation of touch, pain and temperature. The cortical synthesization of touch, pressure, muscle tendon, the effects of contour, form, size and weight, are completely eradicated in the involvement by the tumor mass of the functions which we know reside in the superior parietal lobule. The fact that vibratory sense has remained unchanged would lead one to the conclusion that the representation of vibratory sensibility in the cerebral hemisphere largely resides in the subcortical centers, namely, the thalamus. It is a matter of common knowledge that vibratory stimuli are far more colored by the affective type of sensibility than by the discriminative. The instinctive reaction of children to vibratory stimuli, shown by the withdrawal of the part from the tuning fork, is repeatedly seen, and the same phenomenon is common in adult patients, where the intellectual grade is not very high. The unpleasant character, for example, of the dentist's drill, even in the absence of pain, and the very unpleasant sensations produced by the deep tones in a whistle or in an organ, are matters of common experience. Discriminative recognition of rates of vibration, even very widely separated, is quite difficult and the chief reaction subjectively obtained is, that the deep resonant vibrations are distinctly more unpleasant than the more rapid and less ample vibrations. These facts seem to indicate that the cortical representation of the vibratory sense is very limited and its main end station would then seem to be in the thalamic centers. In none of our cases of pure cortical anesthesia, so far as has been demonstrated at operation in the past six months, has there been any material disturbance in the vibratory sense, and it seems fair to conclude that the major representation of the vibratory sensibility is in the thalamus.

The disturbance in pain and temperature sense consists in a moderate reduction in the acuity of perception to hot and cold and of the sharpness of the pin prick. The hurt element, which we believe to be largely thalamic in origin, is retained; but the cortical contribution in recognition of pain—that is, its location, its sharpness and the promptness with which it is appreciated—are diminished to a slight extent. It seems quite self-evident that there must be some discriminative elements in the appreciation of pain and temperature, and in the disturbance as shown here, there is a fair indication as to what elements of pain and temperature sensibility reside in the cerebral cortex.

In considering the disturbances of sensibility, it is necessary to study the type of aura in this case. This consisted of a pain originating in the lower part of the thorax on the right side and spreading upward to the head and outward to the arm. It is difficult to conceive of any explanation for painful auras in these conditions, but we may perhaps be allowed to consider that the factor, in this case has been some vascular disturbance in the tumor, making itself felt first in the area represented by the middle part of the post-Rolandic convolution upon which the cephalic border of the tumor impinged, and then spreading downward along the post-Rolandic convolution and manifesting itself as a referred pain with its distribution first in the thorax, then in the arm and finally in the head. The fact that the aura consists chiefly in pain may be explained by an excitation of the corticothalamic fibers passing from the post-Rolandic area to the thalamus, in which case the cortical arrangement would present the pattern of the disturbance whereas the thalamus contributes the affective elaboration, namely pain.

DISCUSSION

Dr. McKendree. I have seen this patient before, when presented at the Neurological Institute. As I recall it, I with others was led to localize the lesion somewhat lower down in the postcentral gyrus than was found at operation. I was misled from the accurate localization largely by the nature of the Jacksonian convulsions which the patient suffered. I think this case, with a great many others on record, shows how little value fairly well localized Jacksonian convulsions may have in assisting us in localizing the lesion in the cortex.

One outstanding feature to my mind is the nature of the aura which this patient experienced. Dr. Riley has referred to the pain which was constantly characterized in the same way; pain in the right chest and the right arm. I do not understand that at all, and I simply mention this point to see if any members of the conference have any different explanation from that given by Dr. Riley. I do not understand pain referred from the cortex or from the dura in that locality.

One other question occurred to me; that is whether tumors situated in the cortex, so close to the superior longitudinal sinus, ever give rise to bilateral symptoms. We had one case recently at the Neurological Institute which presented bilateral symptoms. The tumor was located in the frontal lobe largely, and still there was a suggestion of a Babinski on both sides. That tumor was rather extensive, and there was a good deal of intracranial pressure, and I think the consensus of opinion was, that the irritative phenomena on the same side were caused by general increased intracranial pressure. I think one should be on the lookout for bilateral symptoms in tumors which so closely approach the middle line. Of course the superior longitudinal sinus is a pretty dense affair, but it is not inconceivable to my mind that pressure across the midline could cause irritative phenomena.

Dr. Cornwall. My impression was that the tumor was more or less attached to the brain tissue by a small pedicle. I think we would all be interested to know the prognosis of the case and the possible frequency of recurrence where the tumor extends into the brain tissue. I should like to have Dr. Elsberg give us some information in that regard.

Dr. Sands. When we first examined this boy, we suspected a growth in the parietal region close to the Rolandic fissure, because he had a definite change in the stereognostic sense in the fingers. He also had increased deep reflexes. A rapidly growing tumor was suspected in that there was a definite period of onset and a rapid development of symptoms. This was borne out by the subsequent course of the case, as the symptoms developed so rapidly that it was imperative for him to undergo the operation only a month or so after we saw him.

I would like to know the opinion of the conference as to the prognosis in this case. Personally, I feel that it is grave, since it turned out to be an angiosarcoma. The best we can do is to hope that it will not recur in a short time.

Dr. Wechsler. Dr. McKendree's remark about Jacksonian epilepsy is true, I think, only late in the disease. Early in the disease it is a very important localizing sign. The reason why late signs are misleading is that they may be due to edema, intracranial pressure, etc. and are distant, neighboring or secondary in character. So that while it is true that frequently we cannot go by the focal signs, if they are very early symptoms, it is very important to bear in mind for the purpose of localization.

Most neurologists wait for choked discs before they will operate on brain tumor. I am glad that they did not wait for that in this case. To wait for choked disc of advanced

vivo. I think we are too conservative in operating in brain tumors.
degree frequently means doing an autopsy in
I should think that the prognosis would be bad here, in view of the character of the neoplasm.

Dr. Elsberg. One can really divide these tumors into several groups. The first is the typical dural endothelioma. As it grows, it more or less compresses the cortex. When the tumor is removed, one finds the cortex flattened in the cavity in which the tumor lay. This type of growth is usually not very vascular.

The second type of tumor is a well encapsulated sarcoma which arises from the dura, flattens the cortex, and often buries itself in the brain tissue. If the capsule of the tumor is injured, the growth itself will be found to be very vascular.

There is a third type arising from the dura, a sarcoma, also very vascular if it is injured; but the tumor is not well encapsulated. Very often only the portion nearest the dura is distinctly encapsulated. The deeper parts are often not well encapsulated or limited. When this type of growth is removed, the cortex near the most superficial part of the tumor is found to be depressed and flattened, while on the deep surface of the growth, flattened cortex cannot be recognized, and the growth seems to merge gradually into the brain tissue, although a line of cleavage can still be found.

In the patient presented by Dr. Riley, the condition of affairs was that which I have just described, which led us to suspect that this was not an endothelioma, but a sarcoma, as was shown to be the case when it was examined histologically.

Recurrence practically never occurs after the complete removal of tumors of type one and two. In growths which belong to the last group, the chances for recurrence are much greater, as one is never absolutely certain that all tumor tissue has been removed. These patients should have radium or roentgen-ray therapy.

Dr. Strong. It seems to me that this question of pain in a cortical lesion is a very interesting one. The pain cannot be due to meningeal irritation because of its reference. It is not referred to the head, but approximately to the parts represented in the cortical areas affected. I can only account for it theoretically by supposing that the cortical irritation stimulated the thalamus presumably by corticothalamic fibers. The discriminative part is of course performed by the cortex, the affective part produced by the above-mentioned thalamic stimulation.

According to Head's work, where tactile modes of sensibility are most profoundly affected, in cortical lesions, the lesion is more apt to be lower, i. e., in the base of the postcentral and parietal areas, than when the sense of position and movement is most affected. This would apparently create two rather conflicting criteria as to localization, where tactile sensibility is most affected in a lower extremity. One would be in doubt whether to base the localization of the lesion on the segmentary distribution or on the mode of sensibility most involved. In this case the conflict is hardly so pronounced, and I think one would decide on the segmentary basis. Head's further point that when the ulnar portion of the hand is most affected, the lower extremity is apt to be involved, is exemplified here.

Another point, in such cases, is the fact that the centers for the upper and lower extremities appear to be so close, closer than one would expect if one considers the large size of the intervening trunk as compared with the extremities. In other words, the trunk is not nearly so well represented cortically as the extremities, especially the upper. As Head well says, the hand is practically a stalked (cortical) sense organ. This over-representation of certain portions of the body in the cortex is not simply dependent upon cortical differences but upon a greater supply of certain receptors in the "cortical" portions of the body (e. g., tactile corpuscles and muscle spindles) and upon more highly developed cortical pathways (dorsal columns of the cord and medial lemniscus). This is well shown in the distribution, but there is, to me, a puzzling point in regard to the sense of movement. The latter is nearly as accurately judged in the proximal segments of the extremities as in the distal, yet in lesions of the cortical pathway, such as the dorsal columns of the cord, the apical portions of the extremities are usually, I believe, the first and most severely affected. The fact, that the impulses which are used by the cortex have to a considerable extent their own path in the cord, makes it rather difficult to differentiate between a sensory disturbance due to a lesion of the dorsal cord and one due to a cortical lesion, when the sensory disturbance above is used as a basis. There are, to be sure, such circumstances as the thalamic quality of the vibratory mode of sensibility and its consequent presence in cortical sensory loss to help us.

While we thus have "cortical" bodily areas, the fact is not so often stressed that we also have "thalamic" bodily areas and such areas are usually just those areas which are not "cortical." They are especially the trunk and other portions in which tactile and other stimuli (tickling, cold, warmth, pricking, etc.) produce the most marked agreeable or disagreeable sensations. These are the areas to test for thalamic hyperesthesia and this hyperesthesia will often be found to be present in such areas, when it cannot be elicited by stimulation of "cortical" bodily areas. Referring to the question at the beginning of this discussion, i. e., pain in this case, it is interesting in this connection to note that the pain started and was most marked, perhaps, in the trunk and was, I believe, not referred at all to the fingers.

Dr. Casamajor. With regard to the tumor itself: that type of angiosarcoma is seen a good deal in tumors both in the nerve substance and in the dura. Dr. Elsberg has covered the point concerning the type. Endotheliomas simply push the cortex ahead of them and do not invade the cortical tissue. Others from the outer dural surface push the dura in, without invading the nerve tissue. But these angiosarcomas very soon infiltrate and work their way through the arachnoid and impinge upon the pia, infiltrate the pia also and extend down the line of the blood vessels to the cortex.

The prognosis is necessarily poor in this case. While one may make a distinct cut and take out everything in the dura, when it goes to the pia, it is almost impossible to get it out. The result has been very good up to the present and I would certainly favor Dr. Elsberg's suggestion of roentgen-ray in order to prevent, if possible, any further growth.

Dr. Riley. I do not think we can be certain as to the origin of this tumor. Of course the embryological constituents of both the dura and the pia-arachnoid are the same. After a period of contact with the dura by a tumor of this kind arising in the pia mater, it is quite possible for it to have become adherent to the dura mater, so that the fact of its adherence to the dura mater does not necessarily indicate its origin from that membrane.

Dr. McKendree's remark about the possible involvement of the other side I would question, because many patients we see, more or less of a neurotic type, have just as active reflexes as this patient showed on the left side. Even with the spread from the ulnar to the biceps or triceps, I do not think that the reflexes of the left side are pathologically exaggerated. The case that Dr. McKendree quoted, as I remember it, presented a tumor high up in the hemisphere, actually passing over onto the mesial surface of the brain. This tumor was limited to the superior parietal lobule and did not approach the mesial surface by 1 or 2 cm.

The question of the astereognosis has been raised on the ground that muscle joint sensibility was disturbed. My impression of muscle joint sensibility is that it is purely or almost purely a cortical type of sensibility. I do not believe that a lesion limited to the post-Rolandic fissure would materially affect muscle joint sensibility. I think it is only in the area of the superior parietal lobule, where synthesization takes place that disturbances of this type of sensibility have their genesis. Muscle tendon sensibility is primarily a recognition of varying postures and hence it is distinctly of the discriminative or central type of sensory appreciation.

In regard to the aura and Dr. Strong's remarks, two things come to my mind. The extension of the aura was from the thorax to the shoulder into the arm and up into the head. That route would follow exactly the sensory areas as they are arranged in the post-Rolandic area. Given a cause for sudden irritation arising in the tumor, it would spread just as the ripples spread on the surface of a pond when one throws a stone into it. From the anatomical arrangement of those areas, it is possible to suggest that the aura may have arisen in that way. So far as pain arising from stimulation of the cortex itself, I think we are on much less secure ground, because everyone knows that the cortex can be operated upon under local anesthesia, without any appreciation of pain, so that we must consider the thalamus as involved in the disordered reaction. So far as we know, there is not any very discrete distribution of the representation of the body segments in the thalmus, so that it is hard to understand stimulation arising from the cortex going to the thalamus and producing such an orderly successive externalization as we find in this case, unless we implicate the thalamus as the source of the pain and the cortex providing the pattern of the disturbance.

As to Dr. McKendree's remarks about the Jacksonian type of disturbance: The motor manifestations in this case began in the hand. If there was direct cortical pressure resulting from the tumor, the first explosive cortical reaction would be in the leg. We have a jump of 3 or 4 cm. from the location of the tumor in the superior parietal lobule to the location of the motor centers for the arm and hand in the precentral convolution. We know that the main function of the superior parietal lobule is to supply the discriminative forms of sensibility, which reach their acme in the hand and fingers. Consequently the connections of this area, with the area providing for the production of voluntary movement, would be more immediate for the hand than any other part of the body, and it seems perfectly conceivable, that excitation arising in the superior parietal lobule might under certain circumstances, manifest itself by motor disturbances in the area most richly connected with this area of the brain, namely the lower third of the precentral convolution. We know that the sensory apparatus is developed for the purpose of producing motor reactions and it is not doing too great violence to our preconceived ideas to transfer this analogy to the cortex, where irritation of a sensory area could manifest itself in convulsive motor responses. I cannot accept, on account of the anatomical difficulties presented, that direct pressure by the tumor can be held accountable for the production of the hyperkinetic phenomena, and it seems more reasonable to explain them on this basis.

THE PRACTICE OF PSYCHO-ANALYSIS IN A PUBLIC CLINIC*

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A method of therapy, to be practical, must be applicable to patients in all stations of life, since the diseases for which it is intended include all classes. Neither indigence nor affluence is exempt from functional nervous disorder, and the false belief that the psychoneuroses predominate among the rich is fostered by the general misconception of the nature and mechanism of neuroses. The public clinics are crowded daily with patients suffering from all grades of neuroses, from the simplest anxiety states to the most complete conversion types.

Not all of these patients require an elaborate and painstaking analysis for recovery, as is proven by the results of other forms of therapy. All these patients deserve study, if only for scientific interest, with a view of determining the basis of the disorder in the mental sphere. At least as much interest ought to be directed to psychical as to physical states—for the psychogenesis of most of the neuroses needs no defense at present. Yet, unfortunately for various reasons, not as much attention is directed to a complicated personality disorder as to an unimportant and non-pertinent skin lesion.

The feasibility of practicing psychoanalysis in a clinic has frequently been questioned. There are many valid reasons against applying analysis universally, but, when viewed psychanalytically, a better insight of any neurotic complaint is obtained. Such also is the result in certain groups of psychoses.

Let us consider the various conditions dealt with at a neurological clinic.

THE NEUROSIS IN CONSTITUTIONAL PSYCHOPATHIC INFERIORITY

Not always do patients, referred for analysis, approach the intellectual and emotional levels necessary to derive any benefit from this management, or failures in these cases can be sought for, in the nature and mechanism of the simple neurosis. Here the illness is superficial, and its purpose is to conceal an inferior personality. They seek, not treatment, but a postponement of a decision in some problems they cannot solve. They attend to hydrotherapy etc. regularly, though protesting its inefficacy. One then may venture to suspect that they persist in the treatment because it will not cure them. Such a simple neurosis is the patient's solution to inadequacy, and may become a "Career" (Casamajor). Any therapy that nurses the "Career" is more welcome than one that treats the neurosis and solves the problem. Some of these problems are psychiatric and others sociologic, and more often both of these factors are involved. In this class we recognize the Constitutional Psychopathic inferiority of State Hospital designation,[1] and the group neatly termed by Casamajor[2] "The Inadequate." Though the latter term is applied to neuroses in business life, it may properly fit those who are not adequately equipped to meet the responsibilities of marriage. To illustrate:

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*Read at the New York Academy of Medicine, Section of Neurology and Psychiatry, January 10, 1922.

CASE I. Dr. I. S. Wechsler asked me to study a young woman, aged twenty-four, who complained of fainting spells with screaming in public places. He informed me that the patient had had some difficulty with her employer on whom she forced her attentions, and her attacks took place frequently in his presence. She then screamed opprobrious phrases at him in her semi-conscious condition. A few interviews disclosed the following situation: the patient had left her husband soon after marriage five years previously and entered into relations with her employer. This man abused her from the beginning and subjected her to many perversive acts. Lately he had neglected her and hence her conduct anomalies. The patient offered a solution to her problem as follows: "It does not pay to stick to one man; from now on I will do differently." An apparently simple solution for one of pathological make-up.

THE NEUROSIS AS A REACTION TO SPECIAL COMPLICATIONS OF THE IMMIGRANT

A complaint similar to the above may arise in an individual without psychopathic make-up, whose environment has radically changed to an inferior level. Such may result through emigration. Though the problems of the immigrant are manifold, yet there are relatively few neurotic reactions directly traceable to mere change of country, in our vast number of foreign patients. Those who are thus affected have special complications, and their reactions are not unlike those of our native patients when similarly situated. We therefore cannot speak of "immigrant neuroses" as an entity, as we do of "war neuroses."

The problems that result in neurosis are those of the individual, and have to be studied from that angle.

CASE II. Dr. C. A. McKendree referred a woman, aged forty, who complained of fainting, insomnia, attacks of weeping and various dysasthesias of anxiety, since her arrival in this country about ten months previously. In Russia, she was engaged in business, successfully maintaining herself and two daughters during the five years her husband was in New York. On arriving here, she was shocked at the youthful appearance of a clean-shaven husband, who had dropped the orthodox customs and habits of her religion, and replaced them with the meager shop talk of a tailor and no ideals higher than card playing for pleasure. He was a poor wage-earner, and economically she was more handicapped here, than when she was in her own business in Russia, besides she missed the meeting with new faces, and the matching of wits with tradespeople—her main form of social contact. Here her only interests were in the preparation of meals and the care of four rooms in a tenement house. This woman was intellectually superior to her husband and felt, that were she to engage her abilities in business, she could do better than he, and at the same time remove the unjustified domination which he claimed on account of his longer residence in this country. She had not the courage to refuse to be his cook except through an attack of anxiety-hysteria.

THE NEUROSIS WITH MORE COMPLEX MECHANISMS

Mild anxiety states resulting from environmental difficulties do not require extensive analytic therapy. Often but a few interviews disclose the conflict and a solution may then be arrived at. It is quite as important in these instances to know when to curtail the analysis, as when to press boldly forth and persevere until the deepest layers in the psychic structure have been exposed. It is in the severe psychoneuroses that the analyst's technique is most useful. Mere conscious contributions by the patient, no matter how carefully noted, would not disclose the unconscious basis for hysterical or obsessional illness. The following case entailed a search of greater psychic depth, and the unwinding of more complicated mechanisms.

Case iii. Referred by Dr. H. S. Howe. A well-bred young lady, aged twenty-one, had periods of unconsciousness lasting many days, during which she would wander away from home, act queerly, scream and weep. Every few months, during the past three years, she would find herself in a hospital, and would be told that she was found on the street, tearing her hair, etc. She could not recall any of the incidents mentioned to her, but vaguely remembered having had severe pains in the left lower abdominal region. After careful study at the various hospitals, her condition was diagnosed Hysteria. The analysis was not completed but the following was disclosed.

Four years previously the patient's father disappeared from his farm, supposedly with some mental disturbance. The patient was the only child and she was her father's companion in his farming duties and outings of fishing and riding. She acted and felt like a boy, and her father encouraged her boyish attitude, and often told her how disappointed he was at her birth, that she was not a boy. Soon after her father's disappearance, her mother sold the farm, moved to a nearby city and practiced Christian Science nursing. The patient was lost without her father's companionship. Having acted the part of a boy, she attempted to displace her father by assuming his rôle in the home. As she says, "To my mother I was a husband, and if not for my illness, I could be as good a protector as he was." Once having identified herself with her father, she acted accordingly, and emulated him by suddenly leaving home without her mother's knowledge and coming to New York. Her illness began about that time. Of that she says, "Since then I seem to have become a different person. Things, that have happened to me in the past, seem as though they happened to someone else and not to me. Things I wanted then, I have no desire for now. I was so sick at the time I would not have cared if they had cut me into a thousand pieces. It is just a mad desire to get away from everything in this world and myself particularly. I did not trust myself to do the right thing. I make so many mistakes. I should not have come to New York. At the hostess house, when I arrived, they urged me to go to a doctor. They feared I was to become a mother. That is the only reason they know of for a girl coming to this city. I cannot see how they think so. To me passion and physical surrender mean animalism, degradation and sensuality, and ends in death. During my attack I get tense; I see a knife, darkness, daddy, something going very fast and striking my face, mother, something wrong, my dad running a long way,my side hurting terribly."

Her illness may be interpreted as follows: Her libido had been directed completely to her father, and with his disappearance, she withdrew it to herself by identifying herself with him. Successful identification may have been better accomplished by her becoming homosexual, and an attempt at this inversion is manifest in a queer attachment she had formed for a puella publica, whom she met at a hospital while ill. She acted the part of a protector to this woman in many morally hazardous situations. She tried to save her by an attempt to displace the men; a further elaboration of her identification with, and displacement of, her father in relation to her mother. Neither her incestuous craving for father, nor her flight away from it into homosexuality, could be tolerated consciously. Hence her attacks of unconsciousness in which incestuous phantasies predominated.

ANALYSIS IN DIFFERENTIAL DIAGNOSIS

Patients with severe neuroses, such as the obsessional types or the non-transference states (paranoia, etc.), may present on superficial inquiry only some mild anxiety states. The causes of the concealment of the true conditions are manifold. The patient at some previous consultation may have been laughed at for his "silly ideas," hence he becomes more guarded. But the ideas persist and there is an urge to release the tension of the accompanying affect, by some anxiety complaint. Some fear detection of the ideas lest they be considered evidence of insanity; others unconsciously know the meaning of their obsessions and fear that the physician may also know the interpretation. It is remarkable how carefully they will ask whether the physician has ever seen a case like theirs, and is not their condition the only one in the universe—revealing a wish that it be so, in order to remain undetected.

Case IV. Referred by Dr. M. Osnato. Was treated at the Vanderbilt Clinic for several years by the usual physiotherapeutic and medicinal procedures. The patient, a married woman, aged forty, complained of various anxiety symptoms, but never revealed her obsessions until she accidentally said, "If only the thoughts would get away from me." When urged to speak about her thoughts, she showed great embarrassment and would not continue. On analysis the following was obtained: Every moment of her waking state she was obsessed by the thought, "a rat in a cat's mouth." She reacted to it differently at various times, with disgust or with fear. She appreciated the foolishness of her obsession but, in spite of her reasoning and no matter how hard she tried to forget it, the thought was forever there. It had begun three years previously, and she recalled that it was soon after she had read a newspaper item about the maltreatment of Jewish women by Polish soldiers in the war zone. They subjected the women to all sorts of indignities, as making them kiss buttocks and genitals. She reflected how terrible would be her plight were she to be made to do it, and how happy she was that she had left Poland many years ago. The vividness of the news item was before her all that day, and it haunted her. But why should she have been so impressed with this particular news item and especially the identification of herself with those alleged maltreated women? Seven years ago her husband came home late at night from a meeting,somewhat intoxicated. She was suddenly aroused from deep sleep just in time to find him attempting to insert his penis into her mouth. She felt nauseated, could not sleep and was very much upset for several days. She tried to reason with herself that nothing had occurred, but the picture of the attempt persisted. She evaded it by recalling how, many years ago, she had kissed her infant son's genitals and the pleasure she derived from kissing him "all over his body." She succeeded for a while in displacing the attempted perversion of her husband by substituting the latter memory, but unwittingly the picture would change to the present state of that infant, who is now a young man. In rapid succession she would evade now one thought and then another, always ending up by picturing herself kissing her son. After several restless days she forgot all about the incident with her husband. Three years ago when she read the above news item, she unconsciously associated it with this experience of four years previous, but avoided her substituted perverted incestuous phantasies by symbolizing them. Hence the obsession of "a rat in a cat's mouth," and the affect belonging to the original incident was transferred to its symbolized substitute. The obsession promptly disappeared after analysis.

In the obsessional neuroses the conflict is taken care of, in the mental sphere, but in hysteria it is converted into physical symptoms, as in the following:

Case v. Referred by Dr. G. A. Blakeslee. A married woman, aged forty-two, complained of a very troublesome symptom. For two years she had been blinking continuously and much more while on the street, which embarrassed her greatly as she was ashamed of it. She could not account for the beginning of it and had been treated with eyeglasses etc., without improvement.

The analysis disclosed that prior to the appearance of this annoying symptom, the patient while riding on a street car, saw her husband walking with a well dressed woman, when he was supposed to have been at a meeting of his lodge. She got off the street car with the intent of following them, but they turned a corner and she lost sight of them. Her husband came home late that night, and the patient, who is a very quiet, religious woman did not dare to question him about the incident for fear of disturbing their happy domestic relations, but she could not sleep that night. All sorts of ideas relative to his infidelity tortured her, but as he continued to be as attentive to her as before, she decided that she might have been mistaken about what she saw. A few evenings later, she again saw him with the same woman and this time she followed until she saw them enter a theatre. Though convinced that she was not mistaken, she still did not dare to talk to her husband about it for fear of the consequences, the notoriety, and the breaking up of the happy home which she prided herself she had maintained for so many years. To quote the patient: "Really it did not bother me after a few days and I got the whole thing out of my head, especially when I got this terrible blinking, and my husband is all concerned about it and he would do most anything to have me cured." It was quite evident that what the patient did not dare to do consciously, namely calling the attention of her husband to what she saw, she did unconsciously by her symptom, and the blinking protected her also from seeing any more that which she could not tolerate. Though the patient saw the mechanism, the blinking disappeared only after she had had a frank talk with her husband, and the incident had been explained by him as a foolish flirtation.

Apparent environmental difficulties may wrongly be blamed for nervous symptoms, when the cause lies in a disturbance of inner relations of the personality.

Case vi. Referred by Dr. L. Casamajor. The patient, a man, aged thirty-seven, complained of severe pain in his eyes. This began three years ago and a radical Killian operation had been performed by a surgeon who hoped, in spite of negative roentgen-ray findings, that he had remedied the condition. Also a sub-mucous resection had been done without relief. He was obliged to give up his position as a railroad engineer which he had held for many years, as he blamed that for straining his eyes. Concomitant with his eye symptoms, he complained of a compulsion to push people off the subway platform on the tracks when a train was approaching, and of a desire to scream loudly in quiet places. He feared that he was becoming insane. He had recently returned from Ireland, where he had gone with the hope that he might improve in his native land, where he had planned, prior to his illness, to return and settle on a farm. While there he became worse and went to London to be treated, without results.

The analysis disclosed that the patient was the youngest of sixteen children and much neglected by his mother, who was old in his childhood, and showed symptoms of senility: self centering of interests, stubborness, and some dilapidation. She was also alcoholic. He hated his mother and often wanted to strike her, but did not dare on account of her powerful physique. He recalled phantasies of incest mostly of voyeur and sadistic character. He left home at seventeen, emigrated to the United States and took up railroading. He practiced continence for the most part, considering illicit relations a sin, but he postponed marriage until he would some day return to Ireland. It was when he was about to realize his plans, that his symptoms appeared and they were aggravated by his return to his birthplace. He had horrible dreams, as he complained, and they were of frank relations with his mother, resulting in nocturnal emissions, and other dreams of relations with men.

Here was a complete disorganization of the libido into its various components,[3] when the conscious personality attempted to force it away from its original fixation, the mother, and transfer it to an adult choice, a wife. Even this attempted choice, the unconscious directed by causing him to postpone marriage until his return to his birthplace. He was not prepared for this adult reaction, because the unconscious fixations became more intensified, the more the conscious tried to assert itself, and the conflict resulted in an attempt to compromise by forcing the components to take the lead. But that also was intolerable, hence the conversion into their respective negatives, which are the neurotic symptoms: pain in the eyes as the negative for the voyeur component, and fear to harm someone as the negative for the sadistic and homosexual components. It was therefore no accident that the patient, though of poor means, had purchased several pair of expensive binoculars, and had fitted himself with eye glasses though not needing them.

By reflection we may get a glimpse of the causation of the disappointment of one visiting his birthplace; the original seat of the mother image as retained in infantile memories. Of course that image cannot be found by the adult and the search is in vain.

In order to save space I will not quote all the cases studied, which numbered twenty-seven (two of which are reported elsewhere [4]). They consisted of: seven anxiety-hysterias, five conversion hysterias, four obsession or compulsion neuroses, one psychic impotence, six conduct disorders, two cyclothymias and two non-transference cases, one of which was dementia praecox and the other paraphrenia.[5] During the period of two years seventeen of the patients have recovered. Of the others, some are still under treatment, a few have either dropped out or the analysis has been discontinued on account of the nature of the case.

CONCLUSION

It has been shown that in conjunction with other methods for the study and treatment of neurological cases in a clinic, psychanalysis has proven a valuable means in diagnosis and therapy. Though not all cases were brought to a successful finish, for reasons that are evident when reviewing the type of cases presented, insight, not obtainable by other means, was gained in the psychic mechanisms of all cases.

In conclusion I wish to express my appreciation to the staff for their kind interest in referring patients, and to Professor Casamajor, Chief of Clinic, whose hearty cooperation made these studies possible.

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DISCUSSION

DR. WECHSLER. I think that Dr. Lehrman is to be congratulated upon the work he has done. It is something that no psychanalyst has ever done before, that is to treat cases psychanalytically in a clinic. He has proven to us that this is possible. He has frequently solved problems in diagnosis for us with his analyses and at the same time helped the patients. I do not suppose he maintains that he has in every instance followed cases to their psychanalytic conclusions, but he has given us some insight into the mechanism of these cases. The main point, to be emphasized, is that in a limited way psychanalysis can be applied in the clinic.

Dr. Sands. Dr. Lehrman has made a real and valuable contribution in that he has shown that psychanalytic treatment may be administered to clinic patients, without collecting a fee for such treatment. To those of us, who have studied psychanalysis as a means of helping the mentally sick, there have been a few rather puzzling aspects to the technique as usually taught by the older men. For instance, we are told that a fee must be collected before treatment is instituted, that no dispensary patient can be benefited by such treatment, and that the patient must report for an unusually long period of time such as is never heard of in any other treatment. Dr. Lehrman has disproven these methods.

A word about psychanalysis itself. There is no doubt that those who have had the proper preliminary educational and medical training and have devoted themselves to the management of the mentally ill, and who are, at the same time, frank in voicing their sentiments freely and in honestly admitting the limitations of the method, feel that psychanalysis is a real and efficacious means of ameliorating some human misery. Medical men have been skeptical about its usefulness and have for a long time failed to allot to it, its true value, thereby depriving the sick of the benefits derived from such treatment. This offered an opportunity for non-medical men and charlatans to exploit the sick by entering this field, which was neglected by physicians. Thus a vicious cycle was formed, for medical men soon thought it unethical to utilize a method which was so largely in the hands of laymen and charlatans. Fortunately, we are at present beginning to appreciate the necessity for training properly qualified medical men in the use of psychanalysis. No less an authority than Mott, a most eminent neuropathologist, has spoken favorably of this method, as is reported in a recent number of the Journal of the American Medical Association.

While it is true that psychanalysis as a scientific method is utilized and properly so, in the various fields of learning, we must insist that in treatment of sick people, physicians alone should use it.

Dr. Elsberg. Whenever I hear a discussion of this subject, I am mindful of what a wag once said about Homer. He said he did not think that Homer ever existed, but that the poems had been written by another man of the same name. I feel that psychanalysis is dignified by a name, when it really should be called simple common sense. Is it not true, that physicians lacked this common sense for many years, and sent patients with neuroses away, telling them that there was nothing wrong with them?

Dr. Tilney. I do not feel that I should speak on the subject of psychanalysis, because I do not know enough about it by actual experience, but I have seen sufficient of the effects of this study and practice to ask a question, which I believe, decides the point as to whether we should foster, encourage and develop this line of activity. Briefly, this question is: Has psychanalysis been helpful to those who are in need of help? From my personal knowledge, I can very safely answer that in the affirmative. I am sure that it has been helpful. People who suffer have had their conditions improved. Furthermore, it has been helpful in a much more extended and far-reaching sense, in a way, which I think, will bear fruit in the future and the signs of which we are already beginning to see. It is a new means of arousing, and has aroused an interest in ourselves. We are beginning to understand ourselves a little better than we did. Certainly there was no greater stupidity in man than the lack of knowledge of himself, which has existed up to the present time. I believe that psychanalysis is a real step toward the study of personality. Of course it has come in for a great deal of criticism, as almost any pioneer movement is bound to do. Many have not seen its purport at all. I agree with Dr. Elsberg that it is nothing more than common sense, common sense raised to the dignity of conscious recognition on the part of those who have studied personality.

We must put still more study upon that subject than we have before, not only as physicians, but also as trainers of physicians. It is certain that one of our most important future courses will be devoted to the study of personality. I was much interested in a recent talk with Dr. Adolf Meyer, in which he described some of the work he is doing in Johns Hopkins University along these lines. He is giving a personality course to students in the second year. Each student, to be permitted to take an examination, must write an analysis of his own personality and present it to Dr. Meyer or his assistants. From that as a starting point, it is perfectly evident how much advantage the student will gain in understanding the personality of others.

The criticisms that have come to psychanalysis have come, I believe, because this analysis started at the lowest level of personality, the sexual element. This is, after all, an important part of life and as a starting point it offered, perhaps, the most advantageous point of departure into a new field because it could be understood by all, and thus make a broad appeal.

There is, however, some danger in psychanalysis at the present time which should be guarded against particularly, the danger of creating imaginative situations in the patient's mind. The psychanalyst must be careful not to do this. He may easily build up a fable out of the patient's experience and make the patient believe that that fable is in reality his trouble.

We certainly intend to stress psychanalysis as much as we can in the development of our department. We believe that one of the greatest necessities of modern civilization is an efficient means of meeting the increasing number of psychoneuroses. We are keeping psychanalysis clearly in mind, as we are thinking of the future Department of Neurology and Psychiatry in Columbia University.

Dr. Casamajor. The discussion has interested me very much. Dr. Lehrman has taught me one thing which I had always thought was true, that psychanalysis can be practiced in a clinic. A certain amount of analytical work can be done, not in all cases by any means, but in a considerable number. He has also shown me that those psychanalysts, who have pointed out that it is necessary for the patient to pay, or the treatment will not do him any good, are going entirely too far, an example of what Paton calls "wishful thinking." Dr. Lehrman has achieved results which we could not have achieved otherwise in the clinic. I do not think it is of so much importance that only a short time can be devoted to the patients. If you can analyze them enough to enable them to live happily, they are cured. I am of the opinion that any patient who requires eighteen months or two years of analysis, six days out of the week, is probably not worth while, and the end result is not going to be very much. If one has a man who is foolish in his reactions, lacks the element of common sense, one can analyze him eighteen months or two years and when one is through that patient will still be a fool. One cannot inject the gray matter into an individual by analytic procedure or any other.

The question as to who should do analysis has been touched upon. Certainly a man to do analysis should know what he is doing. He should be scientifically honest; he should be careful not to inject his own ideas into a patient, as Dr. Tilney has pointed out. When a brilliant suggestion occurs to him which would give a dramatic explanation to the whole group of symptoms, he must analyze himself and see whether this has come from the patient or from him. Secondly, it should be done by a doctor, not by a layman. Most of the quacks whom we see in psychanalysis belong to the latter group. Furthermore, it should be done by a doctor who is doing something else. He should not restrict himself absolutely to psychanalysis. He should know how to use his percussion hammer and stethoscope. If he does not see organic cases in private practice, he should work in a clinic, keep his feet close to the ground, and realize that the organic side of the nervous system is important. But just because psychanalysis leaves itself open to criticism and lends itself to quackery is no reason why we should turn it down. Endocrinology also lends itself to quackery and yet it is an important field of neurology.

Dr. Elsberg brings up the question of common sense. I should like to ask Dr. Elsberg his definition of common sense. It is a thing which we accept, but do not define. I think it was Molière who said that common sense is the highest attribute of the human mind. It certainly means ability to judge. If an individual lacks common sense, it must be for one of two reasons; one is that he lacks judgment, that is, that he is feebleminded; the other is, that on account of his personal make-up, he cannot bring his judgment to bear upon certain phases of the question. What psychanalysis does is to remove certain prejudices. By hauling out these prejudices into the light of day the individual is able to apply to them every particle of his judgment, so that when the thing occurs again, he does not react in a prejudiced way—lacking in judgment—but in a common sense way. Of course psychanalysis is common sense, and it is gratifying to hear that from a surgeon.

Rowe has pointed out that in the neurosis there is a sort of vicious circle. Primarily the emotions are involved, which affects the involuntary nervous system; that in turn has its effect upon the endocrines; the endocrines in their turn acting upon the nervous system form a vicious circle. It probably makes no difference where one breaks into that circle. The endocrinologist may break in on the endocrine level, someone else may break in on the involuntary nervous system level. We break in on a mental plane. There is more than one mental plane. One can break in by means of religion, by means of hypnosis, or with Christian Science. We, who are interested in analytical procedure, prefer to break into it on the only plane we know; that is, the plane of the intelligence. When emotional things are dragged up to the intelligence plane they are robbed of their effect. If one can rob them of their effect that is all that is necessary. The idea itself is of no harm. It is the false effect that is harmful. So that an application of common sense to the psychoneuroses is what we bring about, applying the intelligence with all the force of human judgment.

Dr. Lehrman (addendum). I was very much interested in hearing the discussion, and wish to add that Dr. A. A. Brill and a few others had practiced psychoanalysis in clinics several years ago. However, I am mindful of the opportunity to have called attention again to the necessity for continuing this procedure in a neurological clinic.

THE VALUE OF PSYCHOMETRIC EXAMINATIONS IN PSYCHIATRIC WORK

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Within the past decade psychology has devoted much of its energy to the development of standardized tests for the measurement of intelligence. The earliest psychometric examination was originated by Binet. In 1908 he arranged a series of tests grouped according to mental age. In 1911 he revised this mental age scale, and later it was still further revised by psychologists in this country, notably by Goddard, Kuhlmann and Terman. The form most frequently used at the present time is the Stanford revision (by Terman), or a combination of the Kuhlmann and Stanford scales. The Kuhlmann-Stanford-Binet ranges from a mental age of three months to an upper limit of eighteen years. The rating is expressed in terms of mental age, or by an intelligence quotient (the I. Q.) which is merely the ratio of the mental age to the chronological age. In adults, the chronological age used in the computation of the I. Q. is arbitrarily fixed at sixteen years, as it is assumed that on the average, mental development ceases at fourteen or sixteen years.

After the perfection of the Binet, other psychometric examinations appeared. Yerkes and Bridges and Hardwick presented a point scale for the measurement of intelligence, which included many tests of the Binet series, but which expressed the rating on the examination in points of credit instead of in mental age. The point scale method failed to supplant the Stanford-Binet, and has gradually fallen into disuse in many clinics. There are, however, several age-level tests which are generally used to supplement the Stanford revision. The Porteus maze tests, standardized at ages from three to fourteen years, and the various performance tests devised by Knox, Healy, Pintner, Patterson and others, are most often used in conjunction with the Stanford-Binet, and are particularly valuable in cases of foreign-born subjects in whom there is language difficulty, with deaf-mutes, etc.

Psychological work in the army gave an immense impetus to psychometric testing and particularly to the perfection of group examinations. The alpha and beta tests, used in estimating the intelligence of the army men, became a starting point for the arrangement of simplified scales for use in schools, and the National Intelligence Tests, the Otis tests, the Haggerty Tests and many other group tests have become familiar to educators. These group examinations offer a fairly accurate method of classifying pupils according to mental age, and segregating them into groups of average, superior and subnormal intelligence, which is of obvious pedagogical advantage, since it permits uniform advance of each group and does not necessitate overworking some children and delaying others for conformity to the average type. The enormous saving of time makes the group tests more practical for educational purposes than the individual tests, although only the latter can be used for diagnostic implications.

The educator has also at his disposal standardized tests in all the school subjects, as well as for the prognosis of his pupils' ability in various fields. The various tests for the detection of special abilities and disabilities are of assistance in the solution of such pedagogical problems as inability to read, to do arithmetical problems, etc. Of equal importance are the tests of vocational aptitude, such as the Stendquist tests of mechanical ability, the Thurstone engineering aptitude tests, etc.

The use of intelligence tests in connection with psychiatric work offers a fairly new field, but one which is rich in opportunities. The psychiatrist is called upon to diagnose and prescribe for many different types of individuals, from the frankly insane to those borderline cases in which it is difficult to determine whether primary amentia or incipient psychosis is the real trouble. Moreover, today the psychiatrist is called upon to pass judgment upon individuals who show aberrant conduct, from the child who plays truant in school to the actual criminal who comes so strongly into conflict with social codes, that he is arraigned before the court which demands of the psychiatrist an opinion as to whether this delinquent is responsible for his conduct and hence is justly liable to punishment, or whether he is irresponsible on account of mental deficiency or mental disease and is therefore to be treated as a person who is mentally sick.

The value of intelligence tests in selecting from the heterogeneous group those who are undoubtedly mentally deficient and have been so from birth is obvious and needs no elaboration. It is worth noting, however, that there are other advantages afforded by the utilization of psychological tests in psychiatric work. Not only the final rating, but also the various reaction patterns to the situations presented in the course of the examination are of diagnostic significance. The psychologist who is experienced in dealing with borderline cases in which an exact diagnosis is difficult, soon learns that certain types of response indicate the probable existence of some mental disorder other than primary deficiency, no matter what the rating on the examination may be. An uneven and erratic distribution of performance on the Stanford-Binet, in which the subject fails on tests which are graded at a fairly low age level but succeeds upon tests graded at a higher level, is always just cause for suspicion of some other factor than simple amentia. A marked delay between the presentation of the problem situation and the response, necessity for constant repetition of instructions in order to secure a response, irrelevancy and inadequacy of the response finally obtained, difficulty in transferring the attention from one problem to the next, reaction to a given situation in harmony with instructions connected with previous situations (i. e., perseveration); all are signs of some interference with the proper functioning of the faculties of attention, association and other mental processes. Cases of incipient dementia praecox are particularly apt to show a combination of some or all of these reactions to the Stanford-Binet. They are also apt to make a much higher rating on the Performance Scale than on the Binet, probably because the performance tests make less severe demands upon attention and association. In manic-depressive psychoses and some other types of mental disorder there are also peculiar reaction patterns and discrepancy between the ratings on different tests. The psychologist who realizes the significance of these factors is able to offer information which is of more material assistance in diagnosis than the mere reporting of an I. Q. or mental age.

The discriminating use of the psychometric examination in those borderline cases in which the psychosis is very mild, so that symptoms are difficult to establish, affords a saving of much time and energy to the psychiatrist, as is well illustrated in the following cases:

Case i. M. K., aged seventeen. A boy is brought to the neurologist by a mother who fears that he is developing delinquent tendencies. He has a history of petty thieving, a poor work record, and is reported to have repeated the eighth grade three times, although previously able to get on well in school. He refuses to work, plays only with small children, etc. The neurologist suspects a case of mental deficiency, and sends the boy for a psychological examination. The psychological report reads as follows:

The rating on the Stanford-Binet gives the boy a mental age of twelve years ten months, I. Q. 80. This rating brings the subject above the level of those classed as mentally deficient in the technical meaning of the term. The performance ranges from a basic year of seven to a group limit of sixteen years, with several failures at the twelve year level, almost complete success at the fourteen year level, and then a sudden cessation of successes with only one correct response at the sixteen year level. This uneven distribution of performance is not in itself sufficiently erratic to excite suspicion, were it not for the fact that it is accompanied by a noticeably indifferent attitude with apparently no affective reactions. This dreaminess and indifference would make one suspect some other factor than simply innate dullness, perhaps an incipient mental disorder of the dementia praecox type.

The boy's history is also suggestive of some mental trouble. He was always seclusive and never played with the other boys, but sat by himself and read. Up to the eighth grade he had been fairly successful in his school work. When called upon to explain why he played with only small children, he replied: "They are not strange children; they are my little cousins." He states that he stole money from his aunt because he wanted to sit in a box seat at the theatre. He appears not to realize the inadequacy of these explanations, which shows a lack of insight on his part. Since the rating on the examination precludes the diagnosis of mental deficiency, his history and behavior must be interpreted as indicative of some developing mental disorder or some personality deficiency.

Case ii. J. S., aged eighteen. Another mother brings to the neurologist her son who has apparently recovered from an attack of manic-depressive psychosis. She wishes to know if the recovery is sufficiently complete for the boy to resume his school work and other duties. The doctor finds little evidence of remaining psychotic symptoms, but calls upon the psychologist for an opinion on the basis of psychological examinations. The psychological report is as follows:

On the Stanford-Binet, the patient makes a mental age of thirteen years. His chronological age is eighteen, giving him an I. Q. of 81. On the short performance scale, the final score is 216. This would be equivalent to a mental age of fourteen years six months, and an I. Q. of 90. It is probable that in neither case does the rating of the test represent the real ability of the subject. His manner of responding to the Binet is significant. His reaction time is exceedingly slow, and constant repetition of instructions is necessary. Unless such repetition is made, the patient either does not respond at all, or responds irrelevantly and inadequately. Such reaction pattern indicates some interference with the processes of attention and association. On the short performance scale, which makes less stringent demands on these faculties, the rating is much higher.

It is to be noted, also, that the rating on these tests is not in harmony with the patient's school history. Just before his illness which began six months ago, he was doing creditable work in the last year of high school. Terman estimates that this requires a mental age of approximately seventeen or eighteen on the Stanford-Binet, while the mental age which the subject is able to attain on the Binet at this time is only thirteen, as already stated. This is another indication that he has not yet regained the complete use of his intellectual functions.

In view of the rating and type of reaction on the psychological examinations, plus the fact that the boy has very little insight into his illness and can remember almost nothing that occurred in that time, one would judge that the patient's recovery is not yet complete, and that it would be unwise for him to resume his full duties at school and in his father's store, although he might be given sufficient work to keep his mind occupied without overtaxing him.

In both these cases, the practical value of the psychological report is obvious. In the latter, it aids materially in determining whether the subject is ready to resume his school work and regular routine. In the former, it rules out the possibility of mental deficiency which is suggested by some of the outstanding features of the case, and enables the physician to concentrate his efforts on determining what factor in the personality make-up or what incipient mental disorder is the real cause of the difficulties. It is just such types of cases in which the physician and psychologist can consult to mutual advantage.

CASE III. R. M., aged forty-three. This was a case in which there was controversy over some property, one side claiming that the subject was a dementia praecox of long standing. In addition to the psychiatric testimony, the results and report of the psychological examination were introduced into the testimony at court. The psychological report read as follows:

On the Stanford-Binet, the subject makes a mental age of seventeen years nine months. His chronological age is forty-three, making the I. Q. 111. This classes him as of normal intelligence. His reactions to the examination are somewhat enlightening. His general attitude is one of anxiety, which is manifest in a tendency to overelaboration of responses, desire for more specific instructions, obvious effort and eagerness to obtain the highest possible degree of success, etc. This anxious attitude is normal in view of his conception of the importance of the test for his case; an attitude of indifference would be the abnormal reaction, indicating lack of insight into the existent circumstances. Moreover, the subject has come for the examination at the end of a day of nervous strain and anxiety in court, and this attitude would tend to hold over. In spite of the mental fatigue which such a day must occasion, his attention and retention is excellent and his comprehension of the situations presented is entirely adequate. This is also strongly indicative of normality. In cases of mental disorder we expect some failure of the processes of attention, association, retention and other mental faculties. The performance on the test is even and regular in distribution. The basic year is twelve and the group limit is eighteen years. In cases of mental disorder, on the contrary, we usually find a decidedly uneven distribution of performance on the Binet, due to lapses of attention and failure of other mental processes.

The subject was finally declared to be of sound mind and awarded the decision in the case, the psychological report being a valuable part of the testimony which won this decision.

Neuropsychiatrists have for the most part become aware of the advantages to be derived from the use of intelligence tests such as the Stanford-Binet and the Performance Scale, both for diagnostic purposes and as an aid in determining how far the patient has recovered from a temporary psychotic attack. In private practice the psychologist is found to be of assistance in such cases as those reported above, while in hospitals for the mentally sick their services are also employed. On active psychopathic services, psychological examinations are given in many instances as a part of the routine for borderline cases and mild or beginning mental disorders, as well as to patients suspected of simple mental deficiency. In hospitals for the insane, the mental tests are used upon patients coming up for parole, to aid in determining whether the subject is able to resume the status in society which he had occupied previous to his mental breakdown.

On the other hand, psychologists have been somewhat slow to realize the necessity for some psychiatric training and experience with psychopathic and borderline subjects as a part of the equipment indispensable to the clinical psychologist. As a matter of fact, data are obtained in work with these types which are exceedingly pertinent to the problems of clinical psychology in other fields. The understanding of the meaning of aberrant reaction patterns, which has already been mentioned, is of undoubted assistance in summing up the clinical picture. In addition to this, the psychologist learns from experience with the types of subjects encountered in a psychiatric clinic that there are certain circumstances in which the rating on a single intelligence test cannot be regarded as reliable. Although the fallibility of the I. Q. was pointed out by Dr. Mateer in connection with the developmental changes in the child's mind, and although Dr. Bronner made an excellent study of the effect of the attitude of the subject on the performance of psychological tests, there has remained a general tendency to accept the I. Q. at its face value, except in instances of evident lack of cooperation or extreme mental disturbance. Experience with a wide variety of cases demonstrates that there are certain definite types of subjects in which the I. Q. can be regarded as only tentative.

The most numerous class in which a fluctuation of the I. Q. is to be observed, is the group which for want of a better term is often designated by the psychiatrist as psychic "constitutional inferiority." Individuals of this type are also characterized as neurotic or emotionally unstable. There are many instances on record in which members of this group have been diagnosed as high grade morons or borderline defectives, on the basis of a single test, when another examination, which chanced to find the patient in a different mood, showed an increase of from 10 to 20 points in the I. Q., and ranked the subject of normal intelligence. Very often in these cases there are no peculiar reactions, no scattering of performance or anything else to rouse suspicion in the mind of even the most highly trained psychologist. It is only through long (and sometimes sad) experience with these unstable types that we come to realize that their intellectual efficiency varies with their mood. Once we have learned this, we are cautious in diagnosing as feebleminded any individual with this unstable make-up unless the I. Q. obtained is at least 20 points below the borderline of normal intelligence, i. e., below 70.[1]

We find also that simple affective disturbances such as fear and anxiety, which are natural emotional reactions under certain situations, tend to interfere with attentive and associative processes, and therefore to prevent efficient intellectual functioning. This affective blocking of the mental processes is apparent in the I. Q., which is materially lowered in these cases. When the cause of fear or worry is removed, and a second examination is given, the I. Q. is apt to be raised from 10 to 20 points. A number of these cases are reported in the article in *Mental Hygiene* already referred to. Our findings in this respect substantiate those reported by Dr. Bronner.

The psychologist on a psychiatric service is often asked to make examinations of children who are convalescing from encephalitis. It will be found that encephalitic patients pass through stages in convalescence which simulate the clinical picture of feeblemindedness very closely. There is an indifferent attitude, poor attention, defective retention, etc. Diagnosis is unwise in these cases, as there is a tendency to regain the efficient use of intellectual functions, so that a later test will sometimes reveal a difference of 20 or 30 points in the I. Q. It is a matter of weeks and months for recuperation of the mental faculties to take place, but the prognosis is favorable, so far as intellectual functions are concerned. It is quite different in the matter of the emotional reactions which do not return to normal functioning with the same facility.

Another class of patients in whom fluctuation of the I. Q. is to be expected are the drug addicts. Here again we find a difference of from 10 to 20 points in the I. Q's, taken before and after the administration of a drug, or immediately after a cure and some time after a cure has been given. To those who are familiar with withdrawal symptoms which appear when the patient is in need of a dose of the drug and in the early stages of a cure, this fluctuation of the I. Q. will occasion little surprise, as the emotional disturbance during withdrawal is sufficiently marked to lead one to expect an interference with attention, association and other mental processes.

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[1] Dr. Jewett and I have reported a number of these cases in the January number of *Mental Hygiene,* in which the first diagnosis of feeblemindedness proved to be incorrect, in an article called "The Influence of affective disturbances on responses to the Stanford-Binet."

In brief, the chief axioms which may be evolved from psychiatric experience in the use of psychometric examinations may be summarized as follows:

1. During a psychosis, an I. Q. is without value as representing the actual capacity of the individual.
2. The I. Q. obtained after a psychotic attack can only be regarded as tentative until sufficient time has elapsed for all emotional disturbance to have subsided and for the patient to have recovered full insight into his condition.
3. In individuals of emotionally unstable make-up, usually designated as cases of psychic constitutional inferiority, two or more tests are indicated when the rating of the first test ranks within 20 points of normal intelligence.
4. In cases in which there is a medical history of organic illness, as epilepsy, encephalitis, neuro-syphilis, etc., the I. Q. cannot be considered an accurate indication of the patient's original intellectual endowment.
5. The I. Qs, obtained on drug addicts are subject to fluctuation of from 10 to 20 points according to the state of the patient with reference to the use of the drug,—whether in withdrawal, recently cured, etc.
6. Even such slight emotional disturbances as the anxiety and fear caused by unusual surroundings may cause an affective blocking which renders the I. Q. taken at the time inaccurate.
7. No I. Q. should be taken as final when there is scattered and uneven distribution of performance, or when any other peculiarities of response such as slow reaction time, necessity of repeating instructions to obtain adequate responses, irrelevant and inadequate responses which may later be corrected,etc., are observed on the Stanford-Binet.
8. When there is undue discrepancy in the ratings on different tests, as in cases in which there is a much higher rating on the Performance Scale, the I. Q. should not be considered final.[1]
9. In cases in which the history, clinical picture, conduct, etc., fail to harmonize with the rating of the test even to a slight degree, a second test is always indicated.
10. The psychometric examination must always be interpreted in the light of all the other findings, and must always be considered in connection with the personal and developmental history, the family background, school history, social reactions, medical examination, etc.

This last point cannot be too strongly emphasized. Even in the diagnosis of cases in which the outstanding features suggest mental deficiency, the psychometric examination is only one of the many points to be considered. The ten fields of inquiry advocated by Dr. Fernald should always be covered in the study of the supposed feeble-minded individual. These include a physical examination, family history, personal and developmental history, school progress, examination in school work, practical knowledge and general information, social history and reactions, economic efficiency, moral reactions, and psychometric examination.[2]

The value of a clear understanding of the principles enumerated above should be apparent when we consider the problems which the clinical psychologist is called upon to handle. The psychologist, who works with criminal and delinquent types in court clinics, meets many psychotics, epileptics, drug addicts, emotionally unstable types, etc. In estimating these subjects, knowledge of the kind outlined in this paper is essential, if incorrect diagnoses are to be kept at a minimum. Even with a broad psychiatric background and experience, the trained psychologist will sometimes be deceived. How much oftener will this be the case if the examiner is deficient in this respect.

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[1] A slightly higher rating on the Performance Scale seems to be customary, but a variation equal to more than a year should be regarded with suspicion.

[2] Fernald, Walter E. Standardized fields of inquiry for clinical studies of borderline defectives. *Mental Hygiene*, April, 1917.

In clinical work in schools, the psychiatric viewpoint is equally essential. In addition to the selection of mentally defective children for special training, vocational study, etc., the psychologist is often asked for advice concerning children who are problems on account of behavior. In many cases, the psychologist who is familiar with psychotic reaction patterns can detect personality defects, which make it imperative that the child be referred to the psychiatrist for preventive treatment, lest a definite psychosis or some other difficulty develop in later years.

Finally, it must be admitted that the chief, as well as the most just, criticism to which the psychologist is subjected by members of the medical profession, is along this same line. The physician feels that the psychologist often attempts to pass judgment upon cases, on the basis of the rating of the intelligence test, without appreciating the importance of a medical and psychiatric examination or the necessity of considering other factors which enter into the picture. The physician realizes that this is due to narrow training and experience; once assured that the psychologist has a general background in neurology and psychiatry and appreciates their importance for study and diagnosis, the critical attitude of the neuropsychiatrist disappears and he welcomes the assistance which the psychologist is able to offer. Thus at the same time that the psychologist increases his own ability by the acquisition of a broader training and experience, he takes a definite step toward the creation of a more harmonious and friendly feeling between the medical and psychological professional groups.

DISCUSSION

Dr. Sands. I think that every medical man, especially one interested in neuro-psychiatry, ought to know about the different tests that are employed in measuring a person's intelligence, not so much with the view of performing these tests but with the ultimate object of being able to interpret the findings in view of the information already gained from the other examinations of the patient. Not all medical men do the Wassermann test on their patient, and yet every medical man should be able to interpret it as it applies to his particular patient.

We should not be discouraged at the multiplicity of the measuring scales, and we must not forget that the findings of the Wassermann test performed at different laboratories are often contradictory, even when we use the blood from the same patient drawn at the same time. The neuropsychiatrist can aid materially in evaluating these tests judged in the light of other findings, such as somatic conditions, emotional reactions, etc. Do we not sometimes disregard the findings of a Wassermann test in the management of the patient?

I am indebted to Dr. Blanchard for her most illuminating talk on a method for diagnosis with which every one who is treating nervous patients should be familiar.

Dr. Tilney. I am very much interested in the problem of mental deficiency. I suppose that, by and large, there is not a factor in human society more potent in the production of unhappiness. One does not have to be in the practice of medicine very long before one realizes this and how widespread it is in almost every community.

I think it is interesting to hear of these exhaustive studies that have been made in rating the various types of defect; but the thing that every time comes back to me is the question, What are you going to do about it? What difference does it make whether the I. Q. is 62 or 70 or 80? That is not actually the problem. It is not helping the poor people who come to us needing our help. The problem is a medical one and ought not to be anything else but a medical problem, regardless of how much the psychologists may be endeavoring at the present time to lift it out of our hands. They are making an error which I think they themselves will see before they get much further in this effort.

We may group the mental defectives into five general classes. These five classes look to the management of mental defectives as an integral part of the social organization. In the first group is the *custodial defect*. The second comprises that large group the *incorrigible but socially viable* defect who can keep his head above water but is in trouble nearly all of the time. Third is the *redeemable defect*, the one for whom we can find some means of management in order to bring him back into a more or less useful sphere in the community. The fourth group is one for which I have borrowed Dr. Walter B. James' designation—the *utility defect,* a type of person whose intelligence attains but a low level, yet one who is indispensable in the servant class. One might mention a fifth group comprising the defects who afford fertile soil for social maladjustments leading to psychoneuroses or going even a step further to the psychoses.

There does not seem very much difference as to the degree of intelligence that the individual may show. We cannot by tests alone tell what should be done in the vast majority of cases. I feel that there are inherent defects in the method of these tests. There is no question that the country child cannot be tested by the same tests used for the city-bred child. Furthermore, there is a constitutional variation from day to day. There is also the important question—which one of us is not defective in some way or other? I was impressed in talking with Dr. Salmon the other day, by a statement which he made to the effect that only 5 per cent of the population of the globe were thinking people. What tests have been brought out to show us the possible variations and defects in the tremendous group of unthinking people?

The observation has been made that the intelligence, insight and general medical knowledge of the examiner are important. To that I would like to add another point; namely, the environment in which the examination is made. If a patient be brought in, tired, at the end of a day in court, what reliance can be placed on the tests? If a patient is in a psychopathic ward, surrounded by psychopaths, influenced by two or three nights' experience in that ward, what reactions necessarily arise in that mind but antagonism and suspicion, and what reliance may then be placed on the tests?

I think by all odds the finest movement in this direction that has been made in this country, is the one started by Pierce Bailey and Sanger Brown in their Classification Clinic. They have given me great help with these problems of defectiveness, more than I have received from any other source. They take a defective child and keep him in good surroundings for about two weeks. The tests are then made slowly, no set time for them is given. The child is studied at his play, at his work; personality studies are made as well as mental tests. The social trends are well worked out. In addition to all this, a complete study is made of the child's endocrinology together with a neurological examination, blood and spinal fluid tests. As a result of this exhaustive study, a report is sent to the physician who has recommended the child to go to the Classification Clinic. On the basis of that report certain recommendations are made. If the child needs motor education, it is suggested. If he is not adaptable to our school methods, that fact is determined and a plan proposed. I had a boy brought to me who had been having epileptic attacks. His mother was concerned because he could not get along in school. She was thoroughly discouraged; his teachers had said that he had a mental defect and was not worth working with in their school—a fine commentary on our educational system. After a complete examination at the Classification Clinic, this fact appeared: The boy had been across the continent during the summer with his parents and had made a collection and a catalogue of all the American wild flowers west of the Rockies. He arranged them in a beautiful manner, cataloguing them with English and Latin names—a really fine piece of work on the part of a boy so young. He was also interested in breeding fine fowl. The clinic suggested taking him out of school and giving him a chance to grow along the lines to which he was especially adapted. He is now being instructed along these lines and was never happier, never better behaved or more easily managed. This, I think, is more important than the intelligence quotient or the exact testing of the mental status; i. e., to find out what we can do for these patients, how we can redeemm or aid them. I have had cases tested by the Classification Clinic and also by other psychologists, and there have been great variations in opinion; so that I have come to the conclusion that there is by no means a constant in the tests even in competent hands. While I feel that the work should be pursued vigorously, the psychologists should bear in mind that they must also show us the means to adapt these abnormal children, so that we may make some use of them and eliminate this prolific source of human unhappiness.

Dr. Casamajor. All of us are born idiots. We come into the world with practically no mental capacity; we cannot take care of ourselves, even utilize what senses we have. I think probably that is what the framers of the Declaration of Independence meant when they said that all men are born free and equal. Starting out, then, from the point of absolute idiocy, the individual develops in two ways, first by elaboration of cortex, the only part of the nervous system that develops in uterine life; second, by adaption to his environment. So that we can speak of real feeble-mindedness in which the defect is more or less inherent in the nervous system, and of another group of individuals, the pseudo-feebleminded, who have no defect in the neural tissues, but who are feebleminded purely from deprivation of the sensorium. For instance, those deaf, dumb and blind, with possibly a perfectly good cerebral cortex. They go through life in a condition of idiocy because they are absolutely deprived of their sensorium. In the case of Helen Keller who, by utilizing a kinesthetic sense in the hands, made contact with her environment, we see how intelligence could be developed, a very high degree of intelligence.

The possibility of deprivation of the environment is quite as important and much more rare. In New Jersey was found a group of children deprived of an environment in which ordinary contacts could be developed, going through life without any social ideals which to us seem to be more or less instinctive, knowing nothing about body cleanliness, who appeared feebleminded, but were capable of education. We might call attention to the historical case of the seventeen-year-old boy, who in 1830 was found in the streets of Nuremberg, standing up practically unable to walk, afraid of the sunlight, thrown into fear at the ringing of a church bell. Taken to a hospital he learned to talk, and inside a couple of years he had reached a level which by modern tests would be an adult level. He had spent his entire life in a cabin in the woods, a cabin so small that he was unable to stand up and had never learned to talk.

Only where we have a definite cortical defect, can we speak of feeble-mindedness.

We can gauge intelligence by mental tests; we cannot gauge the things Dr. Tilney spoke of. Those are conduct things. At present we have no tests to reach conduct. I am skeptical if we ever will be able to reach conduct by means of tests.

The limitations of the tests must be clearly understood, especially by neurologists, I think, if only to overbalance the enormous enthusiasm of some of our psychologists who know nothing of the nervous system and its limitations. We must look upon these tests as very valuable aids in diagnosis. They have been compared to the Wassermann reaction. That comparison is not at all bad. Mental defectiveness is not the sum total of a series of questions and answers; that only helps out in diagnosis—a valuable help but it must be evaluated by other standards. At present a definite skepticism is advisable. If we are going to be able to test intelligence by means of any series, we will have to have a standardized environment of the individual. We do standardize the environment up to the age of fourteen years. All children must go to school. But we cannot standardize the environment from that point on. It makes a great difference, whether from that point the individual goes to work as a messenger boy or as a hod carrier, whether he becomes a lawyer, engineer or doctor. We cannot gauge anything beyond the time when the intellectual environment is standardized.

It is a good thing to see that psychologists are discovering emotions, and realizing that man is not a machine that works on cogs.

The emotional reaction of the individual is most important and must always be evaluated. Until we have some way of approaching that with some degree of accuracy, most tests will be inadequate.

These tests are arranged for mental deficiency, carefully arranged, and probably come as close to perfection as possible, but they cannot be applied to psychoses when the question is one of mental deterioration, not of mental defect. We find this is dementia praecox. Here is not so much an intellectual deterioration as a difficulty in the emotional side of life. More striking than that is the deterioration seen in general paresis. The mind does not deteriorate in levels at all. It deteriorates more in some things than in others.

A final word on how to do these tests. Dr. Glueck told me that any high school graduate with Dr. Terman's book could do a Binet test. I do not believe that any more than I believe that no one can do a Binet test unless he has a degree of Doctor of Philosophy. There are many high school graduates, there are many public school undergraduates, who can make better and more accurate mental examinations than some doctors of philosophy. It requires an individual who can inspire confidence and suggest calmness to the child, one who can win the child's friendship, who has a big measure of common sense, who knows something of the general perspective of children's reactions, not only in their answers but on the emotional side, some contact with general life, something that may reach the child's emotional relations to its home life, personality which is so necessary to a mental test. We do not need doctors for this; most doctors would be very poor at mental testing; psychiatrists possibly would be better than others. It is detail work, and most doctors cannot afford the time any more than they can for Wassermann reactions, urinalyses or blood counts.

MULTIPLE SCLEROSIS IN CHILDREN, WITH A REPORT OF THREE CASES*

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In studying the records of 197 cases of multiple sclerosis, I came across three, which began respectively at seven, eight and thirteen years of age. This interesting occurrence of the disease in children led me to investigate the literature;* but before discussing the subject I should like to record the histories of the three cases.

S. N., school boy, aged 10, born in the United States, was admitted to the Mt. Sinai Hospital in August, 1916, complaining of weakness and paralysis of both legs. He had previously been admitted to the hospital in June 1913, with the history that he had been perfectly well up to the 27th of that month. Birth and development had been normal. One day the mother noticed that the boy's left hand was weak, the next day his right hand became paretic and the following day the boy could not stand up, apparently because of weakness in the legs. The onset was acute, but without fever, vomiting, loss of consciousness or any signs and symptoms of an acute febrile disease. Four days after the onset the boy was admitted to the hospital, where he developed incontinence of urine and feces. The important physical findings were absent abdominal and cremasteric reflexes and the presence of a left Babinski, normal electrical reactions and normal spinal fluid with but four cells. The boy gradually improved, all symptoms disappearing, except spacticity in the left leg.

He was apparently well up to December, 1913, when he began to drag the right leg and developed a tremor of the left hand. He also became somewhat stupid and had laughing spells. He was readmitted February 23, 1914. On examination he showed normal, equal pupils, tremor and ataxia of the left hand and absent lower abdominal and cremasteric reflexes. He was discharged on March 11, 1914, the tremor and ataxia having disappeared. He was again admitted to the hospital on November 22, 1915, when he showed weakness of the left leg, arm and lower face, unequal pupils, right greater than the left, nystagmus, ataxia of the left arm, absent abdominal reflexes, negative spinal fluid and negative blood Wassermann reaction. The boy was discharged in December, 1915 considerably improved and was apparently well to within a few days of his last admission in August, 1916, having gradually developed weakness of the lower extremities. At this time he had equal pupils which reacted normally, horizontal nystagmus to right and left, unequal knee jerks, right greater than left, and bilateral Babinski, more marked on the left side.

From the description of the case, it would seem that the boy had multiple sclerosis. This diagnosis was considered most probable at the time he was in the hospital, although other possibilities were mentioned. The numerous remissions spoke in favor of disseminated sclerosis. Congenital lues was definitely ruled out and so was poliomyelitis. There was no history of cerebral injury either at birth or subsequently and none of infection. The boy had been perfectly well up to the onset of his illness at six and a half or seven years. Hereditary and familial degenerative diseases were also seemingly ruled out. Although there are no pathological findings, it is quite reasonable to accept the diagnosis.

The second case is that of R. Z., a young girl aged eighteen years, of Roumanian parentage, whose illness began at the age of eight. She was in the Montefiore Hospital from October 26, 1915 to May 6, 1916. She had previously been treated in a hospital in Vienna in 1909, in a Philadelphia hospital in 1912 and in the Jewish Hospital of Brooklyn in 1913.

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*A complete discussion of the subject of infantile, congenital, familial and hereditary multiple sclerosis will appear in my article on the Statistics of Multiple Sclerosis.

The past and family history were negative; development was normal. She was perfectly well up to the onset of her illness in 1905, when she began to complain of pains all over the body, especially in the legs. She soon began to drag both legs. This passed away in four weeks, to be followed by weakness and numbness of both hands. Then she found that she could not use her hands as well as hitherto. This cleared up soon, but shortly thereafter she began to experience difficulty in urination and at times had incontinence. Two months later the pains returned and she was confined to bed for six months. Ever since then she had need of the support of a cane. Her chief complaints on admission were inability to walk well, difficulty in control over the bladder and trouble in the use of her hands.

On physical examination, she showed a spastic gait, tremor of the head and hands, spasticity in the lower extremities, greater in the left than the right, lively knee jerks, bilateral ankle clonus, more marked on the left, absent abdominal reflexes, bilateral Babinski, areas of vague temperature disturbance scattered over both lower extremities; some bilateral ptosis of both lids (congenital?) weakness of the right side of the face, some deviation of the tongue to the right, and posterior staphyloma of both fundi due to high myopia. The spinal fluid was negative. The patient was somewhat emotional.

The physical findings apparently warrant the diagnosis of multiple sclerosis. The onset with pains is rather difficult to explain, the suggestion of a hemiplegia less so. The afebrile onset in a perfectly normal and well-developed child and the course of the disease also tend to uphold the diagnosis.

The third case is that of E. I., a young married woman aged twenty-seven, who had been an inmate of the Montefiore Hospital nine years before her readmission and was observed at the Neurological Institute two years before. On admission in August 1920, she complained of tremor of both upper extremities, inability to swallow fluids, slowness of speech, difficulty in walking and contracture of the fingers of the right hand. Her past and family history were negative.

Her present illness dated back to the age of thirteen. It began with shaking of the right upper extremity. The tremor was of the intention type. This was followed one year later by tremor of the left upper and weakness of the left lower limb. One year later the right leg became affected. At the age of twenty, she began to have contracture of the fingers of the right hand and at twenty-one she first experienced difficulty in swallowing liquids. Physical examination showed weakness of the left internal and external recti, pallor of both discs, especially of the right, weakness of both facials, lateral and vertical nystagmus, intention tremor, particularly of the right hand, absent abdominal reflexes, bilateral ankle clonus, more marked on the right, bilateral Babinski and Oppenheim and diminished vibratory sensations in both lower extremities. The blood and spinal fluid Wassermann reactions were negative.

Except for the weakness of both facial reflexes the physical findings leave no doubt as to the diagnosis. It may be added that the same diagnosis was made at the Neurological Institute.

To these three cases I can add a fourth, which began at the age of fourteen. This makes two per cent, of the total number of 197 cases.

In view of the comparative rarity of multiple sclerosis in children and the fact that some people altogether deny its existence at an early age, one must needs show a little trepidation in reporting cases of this group. Oppenheim in 1887 expressed the opinion, in speaking of four cases which he had seen, aged four, seven, twelve and thirteen respectively; "that the beginning of multiple sclerosis in adults can not infrequently be traced to the very earliest childhood." Similarly Muller stated that "proof of the existence of infantile multiple is still missing, but it may trace its beginning to childhood" and "the development of the process on the basis of a congenital predisposition (is) most likely."

Numerous reports of multiple sclerosis in children have appeared in the literature. Marie collected 14 cases up to 1883, Unger 19 up to 1887. Then came Nolda in 1891 and Eichhorst in 1896. The latter added a case of his own, which is one of the very few confirmed by autopsy. Nearly all the rest up to date represent clinical reports. Schlesinger also reported the case of a boy who had subacute multiple sclerosis, which was proved by microscopic study. Schuler's case mentioned by Nolda was also confirmed by autopsy.

Weisenburg described two cases, in brother and sister, of fifteen and four years of age respectively. Stieglitz recorded three cases aged nine, eleven and fifteen, but one is left unconvinced by the description of the cases, although it should be added that he was quite sure that they were true instances. Armand-Delille, Raymond and Beaudouin and Raymond and Lejonne reported cases in children. None of the three is conclusive. Rauschburg cited a case which is said to have begun at one and one half years of age. From the description given the diagnosis seemed not improbable.

A very exhaustive investigation was made by Schupfer. He studied fifty-eight cases recorded in the literature up to 1902 and added one of his own, that of a child of eleven. A perusal of the history of that child leaves one utterly unconvinced. However, Schupfer critically analyzed the material he gathered and decided that twenty-eight of the fifty-nine were authentic instances of multiple sclerosis in children.

In 1909 Gaehlinger reviewed the histories of 86 cases out of a total of 91 hitherto reported in the literature as having occurred in children, including these reported by Schupfer and Baboneix (1904). Gaehlinger concluded that "none of the observations have given absolute proof of the existence of infantile multiple sclerosis." He emphasizes the difficulty of diagnosis and the simulation of the disease by other entities To this may be added, the unreliability of most histories, the unusual difficulty in tracing the onset of the earliest symptoms and the inability to exclude fever or infectious diseases as etiological factors.

From a review of the literature and a critical study of the numerous case histories, one may say that the vast majority of cases stand unproved. In those instances the diagnoses were undoubtedly erroneous; they were atypical forms of other children's diseases of the nervous system, simulating multiple sclerosis. The clinical descriptions (so very few came to autopsy) fit in better with congenital disseminated lues, various traumatic birth palsies, spastic diplegias, Little's or Foerster's types, family spastic paralysis, Friedreich's disease, tumors of the brain, hysteria, encephalo-myelitis and cerebral sclerosis. However there have been a number of well-authenticated cases, which have been proved clinically or pathologically to have been multiple sclerosis. The conclusion, therefore, may be that while the disease is very rare in children, its existence cannot be doubted. But every reported case demands more than usual scrutiny and should arouse not a little suspicion of other diseases of the central nervous system.

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A CASE OF SPINO-CEREBELLAR ATAXIA

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HISTORY

The patient is a negress, aged twenty-three, single; by occupation a switchboard operator. She presented herself at the Vanderbilt Clinic, July 23, 1921, complaining of: (1) Dizziness, (2) inability to walk without aid, (3) headaches, (4) cramps in both legs and right arm, and (5) pains in the back.

Present Illness. The symptoms were first noted four years ago. The onset was gradual. The first symptoms to appear were:

1. "Flashes before her eyes" followed by dizzy sensations. Things seemed to go around from right to left. She would stagger—then everything became dark and she would fall. No headache, nausea or vomiting accompanied these attacks. A sister who witnessed them states that the patient did not lose consciousness nor were any convulsions noted. The duration was twenty to twenty-five minutes. There were no apparent effects.

2. Unsteadiness in locomotion began with the dizzy attacks during which the patient would stagger and sometimes fall. She first noted that it was easier to descend than to ascend stairs. Both legs were affected but the right seemed to be worse than the left. The difficulty in locomotion has grown steadily worse, more especially during the past two years, until, as you see, she cannot walk without aid. The patient says that she never noticed any difference in walking in the light or dark.

3. Headaches began two years ago. They are nocturnal and diurnal. They occur suddenly and are worse at night but do not keep her awake. The frequency is about twice a week. They are sometimes accompanied by nausea and violent vomiting. They have never been associated with the attacks of dizziness. The pain is described as general in distribution over the head with possibly more tendency for frontal localization. They may come on at night and be present when she awakens.

4. The cramps in the legs and arm began with the other symptoms and are growing worse. The muscles get stiff and hard and painful. The cramps are intermittent and occur sometimes as frequently as every night. The duration is about twenty to twenty-five minutes. They affect one or both

legs but only the right arm. The pains are sharp and shooting in character. Her fingers sometimes cramp when she braids her hair.

5. The pains in the back are intermittent, and shooting in character. They are not getting worse. She complains of a tired feeling in the back.

Birth and Development. No reliable history can be obtained concerning the birth or development. She began school at the age of seven and stopped at the fifth grade at the age of twelve.

Injuries, Diseases, etc. There is no history of injuries or of diseases that pertains to the present condition. The vision and hearing are grossly normal. There are no sensory or visceral disturbances except as noted above. There are no abnormal mental or emotional disturbances.

Habits and General Physiological Status. Tea and coffee are indulged in moderately. Alcohol and drugs are not used. She sleeps well. The appetite is good and the bowels are regular. The weight is 119 pounds and there has been no loss. There are no urinary disturbances. Menstruation began at the age of thirteen and has been normal as to interval and duration. There is dysmenorrhea the first day.

Family History. The patient was the eighth child in a family of eleven. Three sisters and three brothers are living and well, their ages ranging from fourteen to twenty-nine years. Two sisters and two brothers died from unknown causes at the following ages: infancy, fourteen, seventeen and twenty-two years. The patient and her sister give the information that their mother had several miscarriages. The father died from accidental trauma. The mother is living and well.

PHYSICAL EXAMINATION

Voluntary Motor System—Gait. The patient cannot stand or walk alone. With assistance the gait is ataxic in character. There is no apparent disturbance of the normal associated movements of the trunk, arms and head.

Coordination. With her eyes open, she cannot stand with her feet together. Standing is only possible when her feet are separated giving a wide supporting base. There is but slight difference with her eyes closed, although the attempt may be less successful. There is a tendency to fall to the left. She cannot stand on either foot alone.

The finger to nose, finger to finger and finger to thumb tests are well performed with the eyes open and closed. There is but slight if any evidence of ataxia but the performance of these acts is accompanied by some tremor, especially on the left, of dyssynergic character. There is a very slight tendency to pass point to the left when the extended arms are tested vertically. This is slightly more marked in the left than in the right arm. In the recumbent position the toe to object, heel, knee and shin tests are well performed with the eyes open and closed but there is evidence of dyssynergia on the left.

Adiadochokinesia and dysmetria are interpreted as absent but dyssynergia present to a slight extent in the arms and legs, more especially on the left.

Skilled Acts. There is no evidence of apraxia or dyspraxia. Speech is normal.

Abnormal Involuntary Movements. There are some medium to fine tremors of the upper extremities and fine tremors of the thumbs. No other abnormal involuntary movements are noted.

REFLEXES

Deep

	L	R		L	R
Jaw	0	0	Suprapatellar	0	0
Pectoral	1	0	Patellar		
Biceps	1	0	Faint response on reënforcement.		
Triceps	1	1	Achilles		
Radial	0	0	Ankle clonus	0	0
Ulnar	0	0			
Wrist	0	0			

Superficial

	L	R
Ciliospinal	2	2
Supraumbilical	2	2
Suprapubic	2	2
Upper lateral abdominal	2	2
Lower lateral abdominal	2	2

Plantar flexion	o	o
Babinski	o	o
Chaddock	o	o
Oppenheim	o	o
Gordon	o	o
Schafer	o	o

Muscle Strength and Status. The acts of rising from recumbent to sitting and standing positions are well performed. Muscle strength of the forearms, thighs, legs and feet is normal.

There is no atrophy or hypertrophy of the musculature of the body. The consistency of the muscles is normal. With the exception of hyperextensibility of the fingers the muscle tone is considered normal. There is no increased muscle irritability or myoidema.

Abnormal Associated Movements and Nerve Status. No abnormal associated movements are elicited. There is no tenderness along the large nerve trunk.

General Sensory. The sensations of touch as tested with cotton wool, superficial pain to pin prick, and temperature discrimination for cool and warm are normal over the face, forearms, hands, fingers, trunk, thighs, legs, feet and toes. Touch discrimination as tested with two points on the fingers is normal (3-4 mm.) There is a doubtful or slight impairment of the vibratory sense in the fingers and toes, more marked on the left. The deep pressure and muscle tendon senses are normal in arms, forearms, hands, fingers, trunk, thighs, legs, feet and toes, with the possible exception of the muscle tendon sense which shows possibly very slight impairment in the great toes.

Cranial Nerves. The olfactory nerve is normal. Vision is normal. The fundi were reported normal by the Department of Ophthalmology. The pupils are equal, 5mm. in diameter, round, central in location, and react equally to direct and indirect light and convergence. Hippus is present. There is slight outward limitation of movement in both eyes. On looking to the right and left a slow nystagmus of moderate amplitude, mostly horizontal but with a rotary component is present. This is slightly more rapid on looking to the left.

The cochlear branch of the eighth nerve is normal. The vestibular apparatus, as tested with rotatory tests, is normal.

No abnormalities are noted in the fifth, seventh, ninth, tenth, eleventh or twelfth cranial nerves.

MENTAL AND SYSTEMIC

The mental status and general physical examination are negative except for the presence, over the face and trunk, of scars of an old eruption, possibly acne.

SEROLOGY

On admission the blood Wassermann test was negative and five days later the spinal fluid was as follows:

Color	normal
Pressure	8 mm.
Cells	1
Globulin	normal
Wassermann	negative with 2 c.c.
Colloidal gold	2223110000.

On July 29, 1921, a course of intramuscular injections of bichloride of mercury was instituted. On August 16, 1921, the blood Wassermann reaction was 2 plus with alcohol antigen and 4 plus with cholesterin antigen. On October 22, 1921, after 13 injections of mercury the blood Wassermann reaction was 4 plus with all antigens. This is a striking example of the provocative value of antisyphilitic therapy. On November 1, 1921, the spinal fluid findings were as follows:

Color	normal
Pressure	8 mm.
Cells	10
Globulin	normal
Wassermann	negative with 2 c.c.
Colloidal gold	2211000000

The serological results warrant the diagnosis of syphilis. The spinal fluid serology is essentially negative.

SUMMARY

To summarize, this patient has practically absent deep reflexes in the lower extremities with greatly diminished deep reflexes in the upper extremities; equilibratory ataxia; dyssynergia; nystagmus; no disturbance of cutaneous sensation, very slight or doubtful impairment of the vibratory and muscle joint tendon senses in the most distal parts of both extremities; normal mentality; normal pupillary reactions; no cranial nerve involvement; no evidence of vestibular involvement; absence of pyramidal tract signs.

The points that present themselves for discussion are:

1. The anatomic location of the lesion.
2. The etiology of the lesion.
3. The pathology of the lesion.
4. The clinical classification and nomenclature.

1. *Anatomic Location.* The bilateral and symmetrical distribution of the symptoms points to a cord lesion but the nystagmus remains unexplained.

The complete absenie of superficial sensory disturbance and the meager or doubtful evidence of disturbance of deep sensation are against peripheral nerve, spinal ganglia or dorsal root lesions. Such location would mean an unusual selective character, because practically only the afferent fibers concerned with the direct reflex arc and spino-cerebellar pathway are affected.

The absence of amyotrophy obviates lesion of the anterior horns. If the lesion is in the cord the whole picture points to a syndrome of the white matter. Our usual conception of myelitis is too comprehensive to fit this picture in the absence of pyramidal tract signs.

There is a locality in the white matter of the cord, a lesion of which would account, I believe, for the major symptoms presented, namely, the equilibratory ataxia and assynergia. This locality is the circumferential zone of the cord, which is traversed by the dorsal and ventral spino-cerebellar tracts carrying afferent impulses to the vermis of the cerebellum and the deitero-spinal and olivo-spinal efferent cerebellar pathways. All of these constitute pathways that have to do primarily with equilibration. A degeneration of the circumferential zone is not at all impossible. Lesions in the brain producing bilateral or symmetrical symptoms are usually bilateral, but a small lesion in the vermis might account for the dyssynergic and equilibratory defects. This would make the ataxia motor in variety, but the impairment of deep reflexes is still unexplained. There are certain points in the serology that favor this location.

2. *Etiology.* Syphilis being proven in this case, I consider it justifiable to regard this as the cause in the absence of strong evidence to the contrary.

3. *Pathology.* The serology suggests the conclusion that the neurologic lesion is not meningeal origin. The absence of pleocytosis or of increased globulin, the practically negative colloidal gold reaction, and two negative Wassermann reactions with the maximum amounts of fluid are inconsistent with the usual serologic findings in meningeal syphilis unless it is very chronic or latent. After the acute stage of meningeal syphilis has passed and the lesion ceases to progress or becomes latent the serology may be entirely negative. The time required for the pathological process to become quiescent, especially when no treatment is administered, is usually longer than the period that has elapsed from the onset of the first symptom in this case to the present.

The parenchymatous or degenerative types of neurosyphilis, usually have certain changes in the spinal fluid which reflect themselves in the colloidal gold reaction giving the so-called luetic or paretic curves, the globulin is increased and the Wassermann reaction is usually positive in high dilutions.

Such serological findings as have been obtained in this case suggest vascular lesions or gummata. It is difficult to conceive of vascular lesions of the cord that could produce a circumferential sclerosis. A meningeal lesion could and frequently does extend to the periphery of the cord but there is no serologic or clinical evidence of meningeal involvement. A vermis lesion on the other hand, does not account for the absent deep reflexes.

4. *Clinical Classification.* Friedreich's ataxia cannot be disregarded in a differential diagnosis. Against this is the absence of familial or heredity history, dorsal column sclerosis, pyramidal tract signs, general sensory disturbances, and pes cavus.

If we assume that this clinical picture is the result of congenital syphilis it is not necessary to limit the lesions to circumscribed areas. The vascular system may have been inherently poor as a result of which thrombotic lesions with resultant softening may have occurred, or there may have been inherent germ plasm defects that are now manifesting themselves. It may be possible that the essential lesions are in the dorsal root ganglia affecting only certain afferent fibers due to a selective activity of the syphilitic virus or to germ plasm defects in these components.

The clinical term spino-cerebellar ataxia is applicable to this case and it may be of hereditary nature, but to call it Friedreich's ataxia compels one to base the diagnosis more on the absence than on the presence of the essential clinical features of this syndrome. This again emphasizes the futility of a diagnostic nomenclature which utilizes the names of clinical observers to the exclusion of the appropriate anatomic or pathologic terminology.

DISCUSSION

Dr. Aronson. It struck me that outside of syphilis, this looks like a Friedreich's ataxia, or an extension of the process beyond the spinal cord as in Marie's ataxia.

This patient has definite involvement of the posterior columns, definite loss of vibratory sense, loss of postural sense. She has nystagmus—that small amount of clinical evidence which one finds in Marie's and does not get in hereditary ataxia of spinal origin.

Dr. McKendree. I am impressed by the presentation of the symptoms in such a way as to make me believe that there is a very diffuse cerebrospinal involvement. It seems to me that there is very clear evidence of posterior column disturbance. The vibratory sensibility is in question. Dr. Cornwall states that he did not obtain equality of response in the toes. Certainly there is definite diminution. With reference to the deep reflexes I think we must consider them pathological, and I do not see how we can attribute the extent of diminution to any cerebellar involvement. I have never seen such a profound disturbance in the reflexes from any purely cerebellar lesion.

With reference to the clinical history, it seems to me that the patient presents evidence that the cerebellum is certainly irritated, and in the early part of her history she had indications of irritation of the lateral lobes: flashes of light, giddiness, intense headache, and vomiting, probably of the projectile type. I believe that we are dealing with a leptomeningitis of the lateral lobes of the cerebellum as well as with involvement of the posterior columns of the cord.

She came in with negative serology; under treatment she has developed positive findings in the blood, although I believe the spinal fluid findings are still within normal limits. Personally, I feel that we are still placing too much reliance on serological findings. There is an interesting article by H. C. Solomon on Neurosyphilis in the November 26, 1921, issue of the *Journal of the American Medical Association,* in which he recounts numbers of cases of definite syphilis of the central nervous system without spinal fluid changes. I think lues is the fundamental factor here; whether congenital or acquired it is difficult to state.

Dr. Wechsler. There is no ataxia at all when she lies down, or at least very little. While I do think that this is an organic case, her gait reminds one a little of astasia abasia, such as you see in functional cases. Her gait is not truly ataxic. She takes no interest in where she is going. I think it advisable to see what could be brought out in respect to the functional side. The absence of pupillary changes, the originally negative serology, throw doubt on the diagnosis of syphilis. A diffuse degenerative condition I think is more likely.

Dr. Tilney. I think the diagnosis is diffuse meningovascular neurosyphilis. It is probably a diffuse scattered lesion, possibly a gummatosis.

Dr. Casamajor. I think this is a diffuse lesion, though Dr. Wechsler's suggestion should not be overlooked. My first impression was that there was a large psychoneurotic element in the case. Today when I saw her walk the idea of astasia abasia occurred to me too, but I threw it out of mind until Dr. Wechsler revived it again. There is no reason why a meningovascular syphilis patient cannot also develop an hysterical manifestation.

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